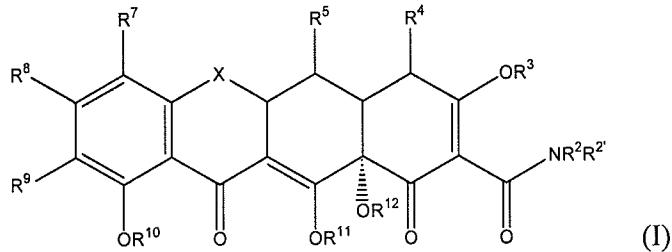


LISTING OF THE CLAIMS:

Listing of Claims:

This listing of claims will replace all prior versions of the claims and listing of the claims in the application. No claim amendments are made herein.

1. **(Previously Presented)** A method for treating a subject for a disease treatable by modulation of RNA (DTMR) associated with splicing of nuclear RNA, comprising: administering to said subject an effective amount of a tetracycline compound of formula (I):



wherein

R², R^{2'}, R^{4'}, and R^{4''} are each independently hydrogen or alkyl;

R³, R¹⁰, R¹¹ and R¹² are each hydrogen;

R⁴ is NR^{4'}R^{4''};

R⁵ is hydrogen;

R⁶ and R^{6'} are each hydrogen;

R⁷ is substituted alkenyl, substituted alkynyl, substituted phenyl, substituted or unsubstituted furanyl, acyl, or aminoalkyl;

R⁸ is hydrogen;

R⁹ is hydrogen; and

X is CR^{6'}R⁶;

or a pharmaceutically acceptable salt, ester or enantiomer thereof;

such that said DTMR associated with splicing of nuclear RNA is treated, wherein said DTMR associated with splicing of nuclear RNA is spinal muscular atrophy, and further

wherein said effective amount is effective to modulate splicing of said subject's nuclear RNA.

2.-36. **(Canceled)**

37. **(Previously Presented)** The method of claim 1, wherein R² and R^{2'} are each hydrogen and R⁴ and R^{4'} are each methyl.

38. **(Canceled)**

39. **(Previously Presented)** The method of claim 37, wherein R⁷ is substituted or unsubstituted furanyl.

40. **(Previously Presented)** The method of claim 37, wherein R⁷ is substituted phenyl.

41. **(Canceled)**

42. **(Previously Presented)** The method of claim 40, wherein said substituted phenyl is substituted with one or more substituents and further wherein said substituents are each independently alkyl, alkenyl, alkynyl, halogen, hydroxyl, alkylcarbonyloxy, arylcarbonyloxy, alkoxy carbonyloxy, aryloxycarbonyloxy, carboxylate, alkylcarbonyl, arylcarbonyl, alkoxy carbonyl, aminocarbonyl, alkylaminocarbonyl, dialkylaminocarbonyl, alkylthiocarbonyl, alkoxy, phosphate, phosphonato, phosphinato, cyano, amino, acylamino, amidino, imino, sulfhydryl, alkylthio, arylthio, thiocarboxylate, sulfates, alkylsulfinyl, sulfonato, sulfamoyl, sulfonamido, nitro, trifluoromethyl, cyano, azido, heterocyclyl, alkylaryl, aryl or heterocyclic moiety.

43. **(Previously Presented)** The method of claim 37, wherein R⁷ is substituted alkenyl.

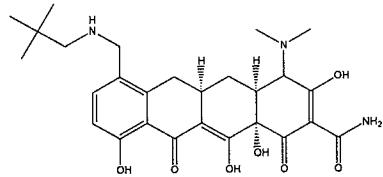
44. **(Previously Presented)** The method of claim 37, wherein R⁷ is substituted alkynyl.

45. **(Previously Presented)** The method of claim 37, wherein R⁷ is acyl.

46. **(Previously Presented)** The method of claim 37, wherein R⁷ is aminoalkyl.

47.-56. **(Canceled)**

57. **(Previously Presented)** The method of claim 1, wherein said tetracycline compound is:



, or a pharmaceutically acceptable salt thereof.

58. **(Canceled)**

59. **(Previously Presented)** The method of claim 1, wherein said modulation of splicing increases splicing of RNA.

60. **(Previously Presented)** The method of claim 1, wherein said modulation of splicing decreases splicing of RNA.

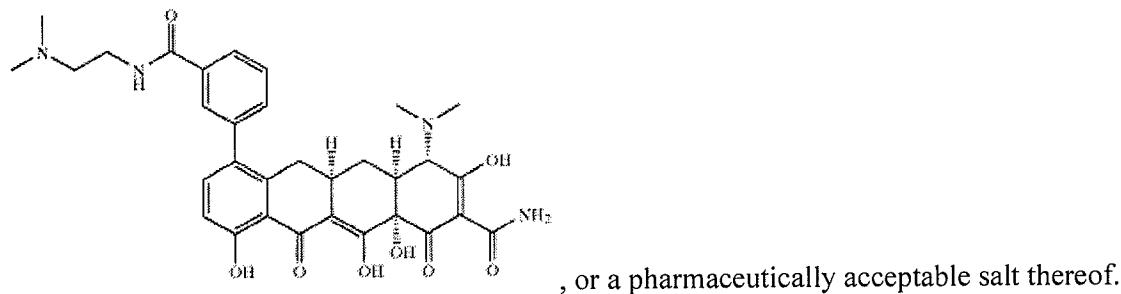
61. **(Canceled)**

62. **(Previously Presented)** The method of claim 1, wherein said subject is a mammal.

63. **(Previously Presented)** The method of claim 62, wherein said mammal is a human.

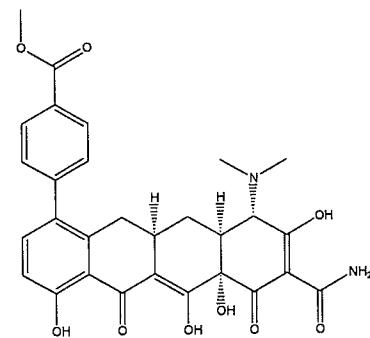
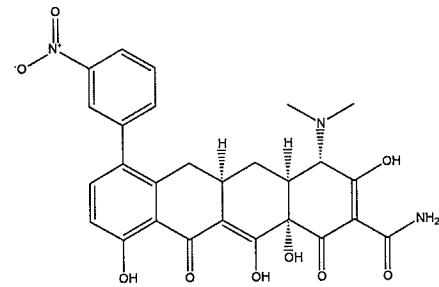
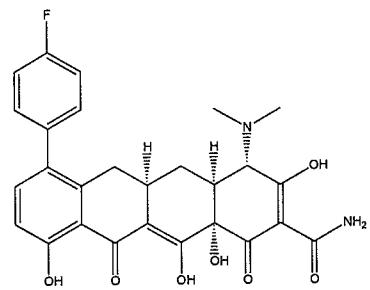
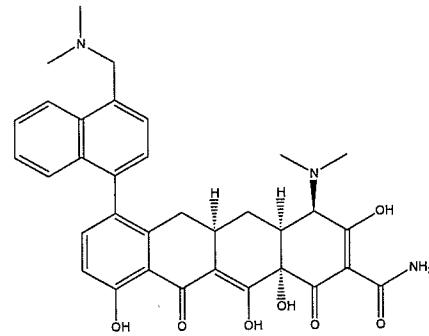
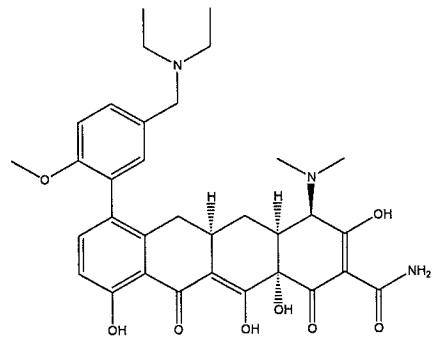
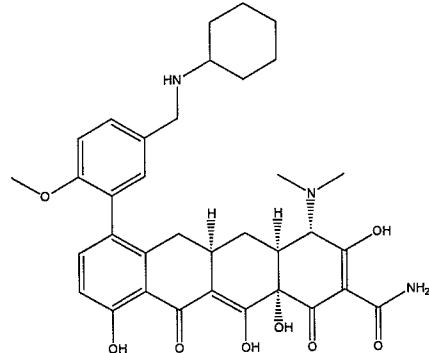
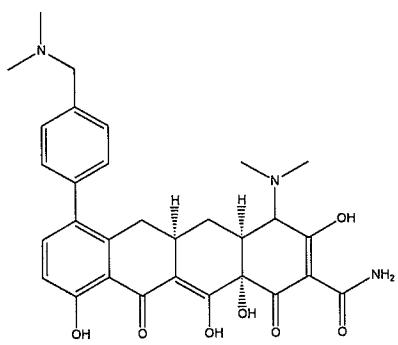
64. **(Previously Presented)** The method of claim 1, wherein said modulation of splicing is activation of cryptic splice sites, silencing of consensus splice sites, silencing of exonic or intronic splicing enhancers (ESEs or ISEs), silencing of exonic or intronic splicing silencers (ESSs or ISSs), alteration of the binding or a component of the splicing machinery to the RNA, or the affecting of intermolecular interactions between components of the splicing machinery.

65. **(Previously Presented)** The method of claim 1, wherein said tetracycline compound is:



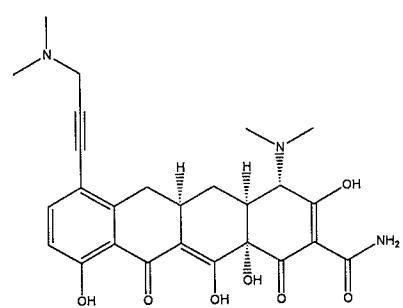
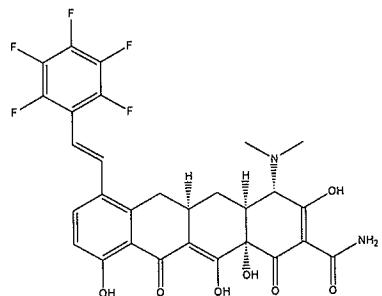
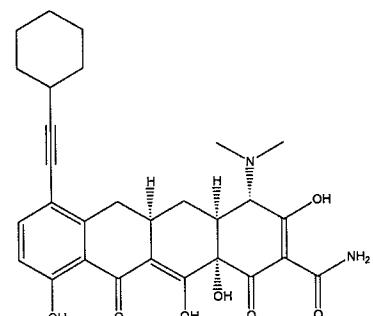
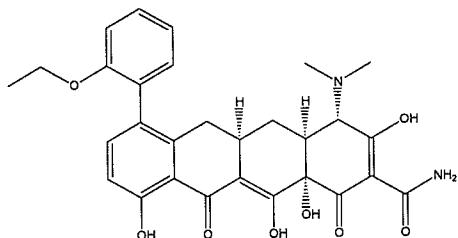
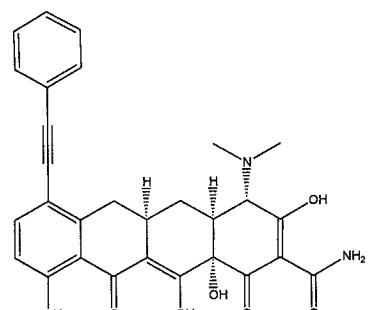
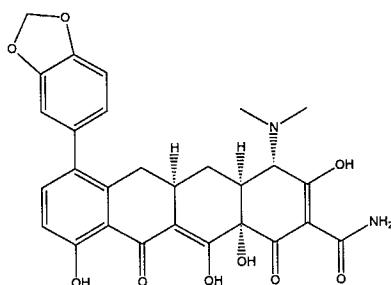
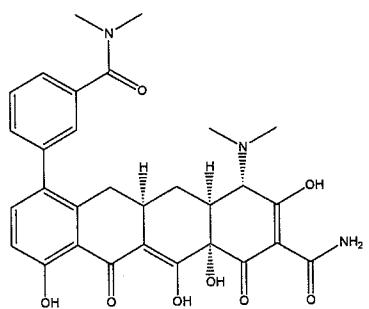
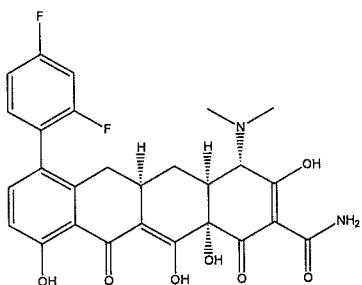
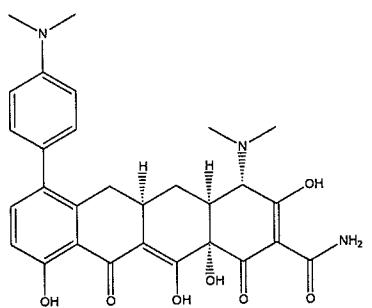
66. **(Previously Presented)** A method for treating a subject for a DTMR associated with splicing of nuclear RNA, comprising: administering to said subject an

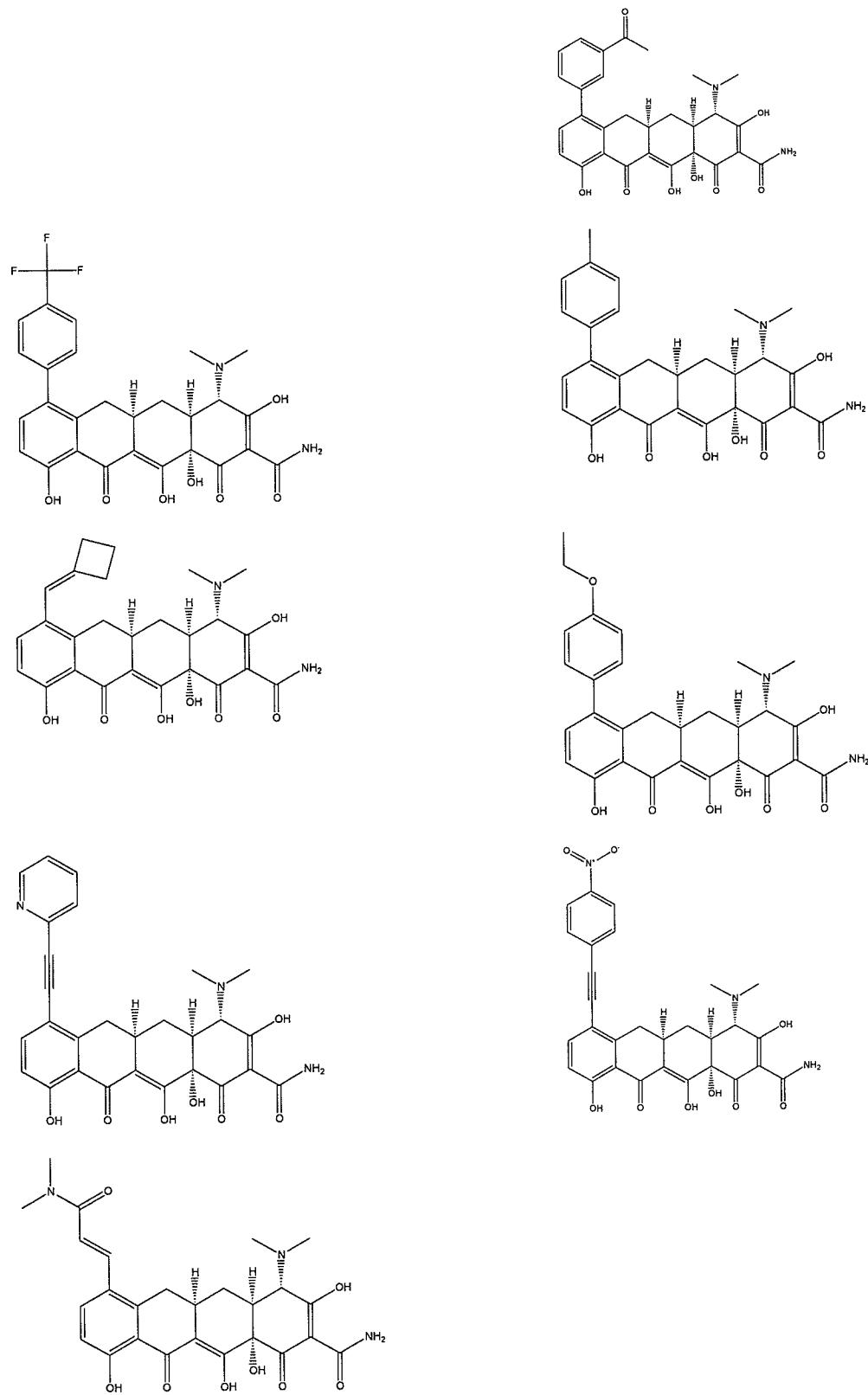
effective amount of a tetracycline compound; wherein said tetracycline compound is a tetracycline compound selected from the group consisting of:

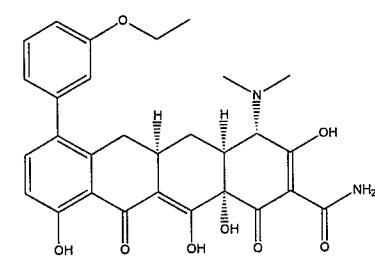
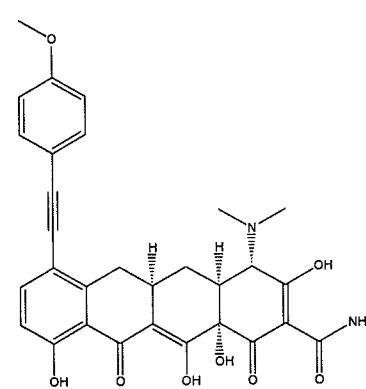
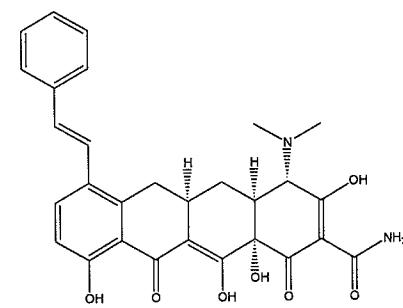
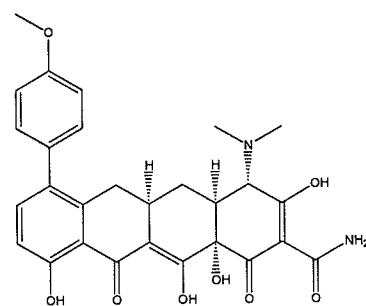
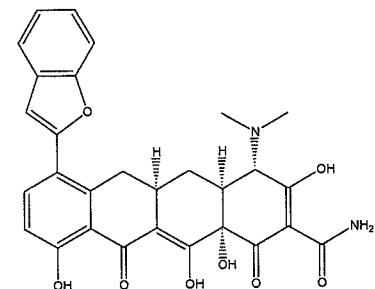
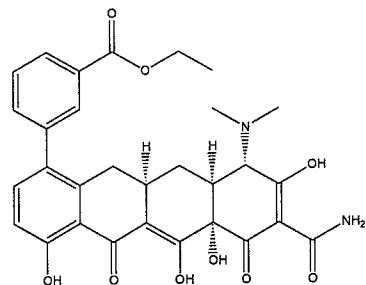
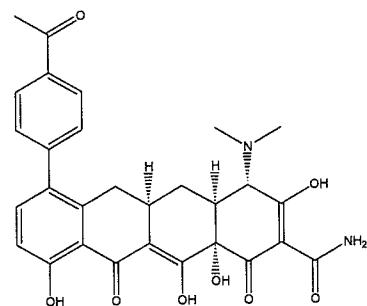
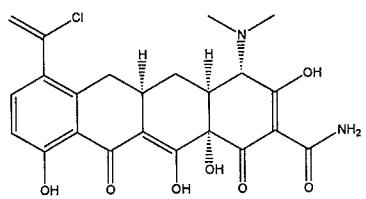


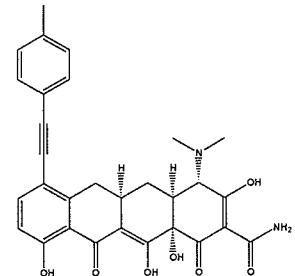
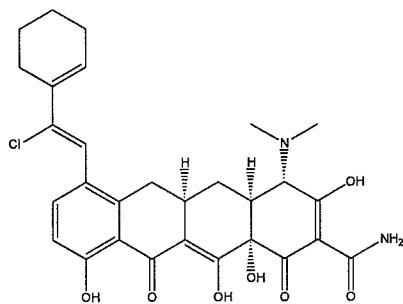
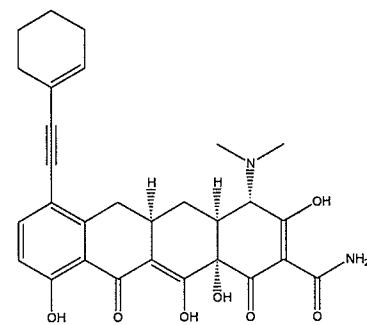
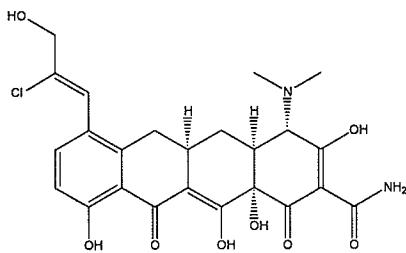
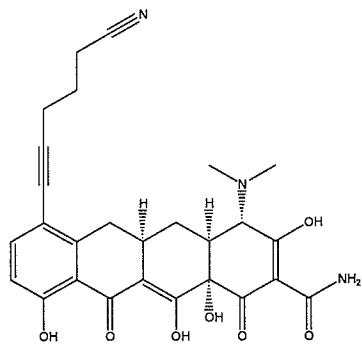
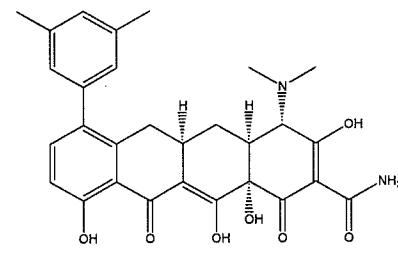
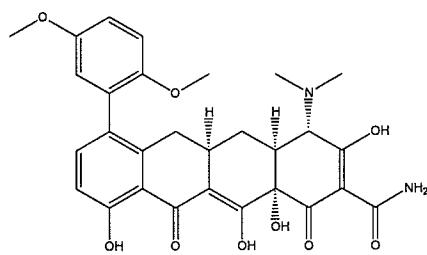
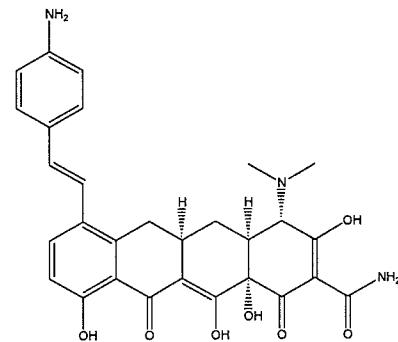
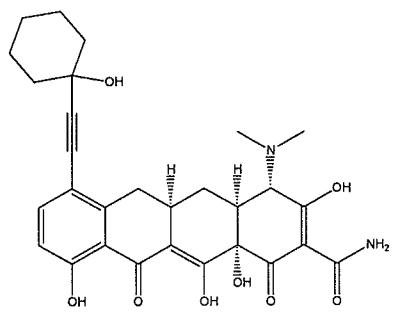
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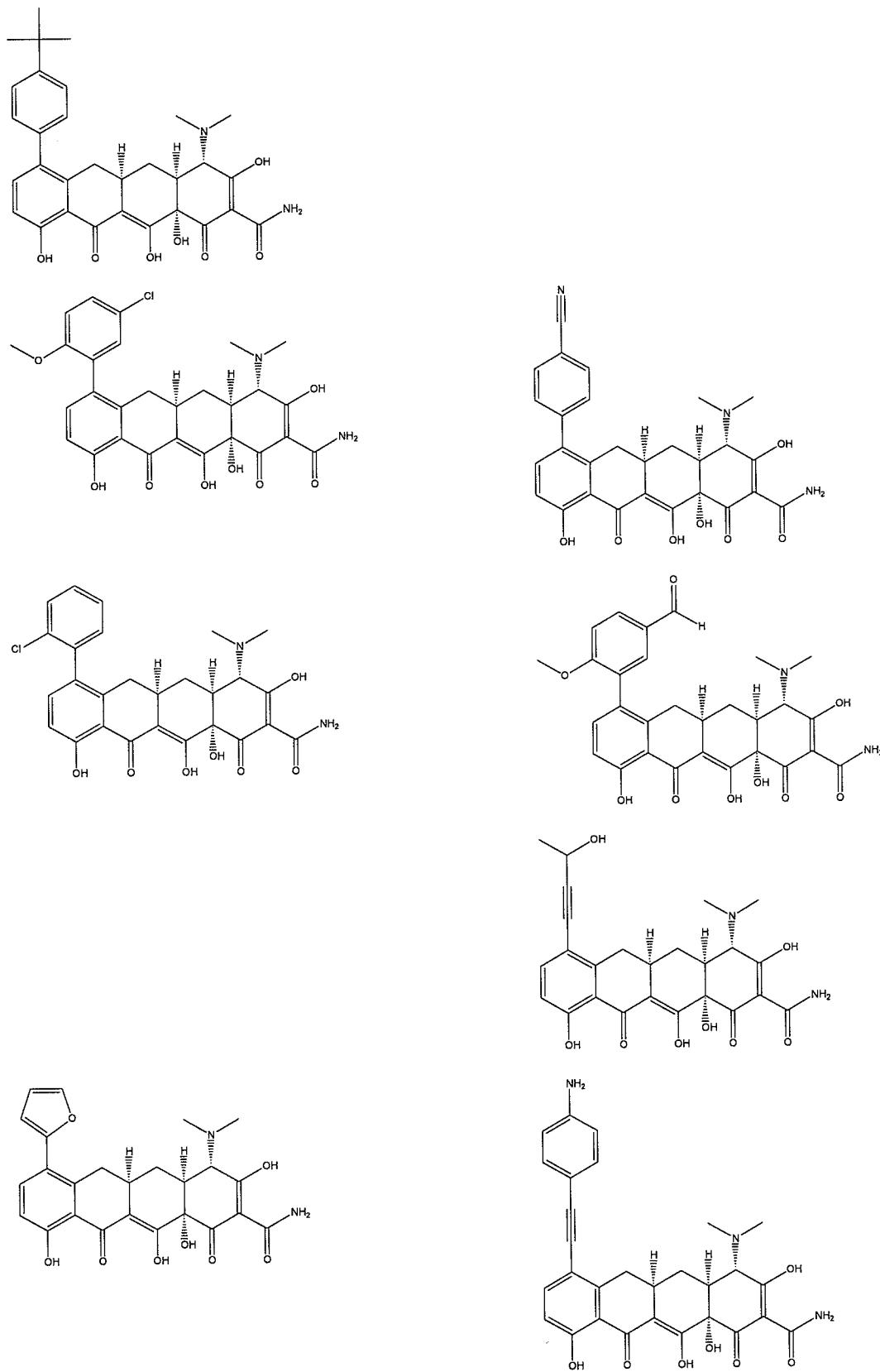
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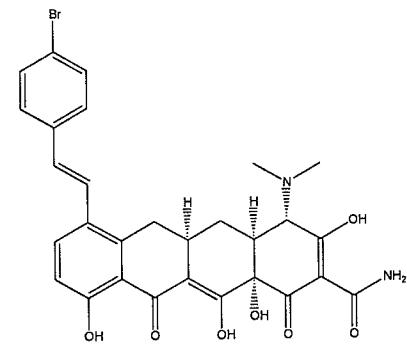
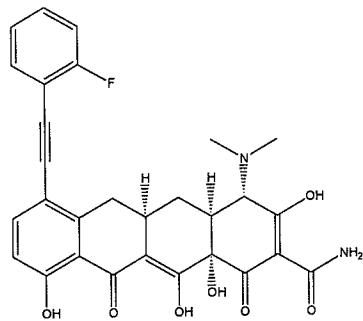
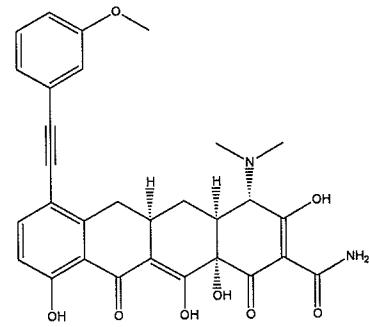
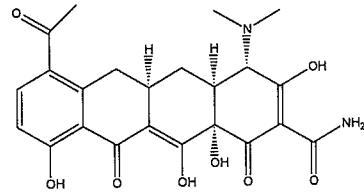
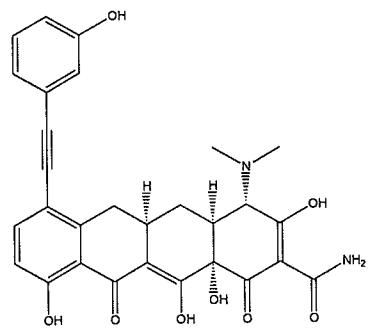
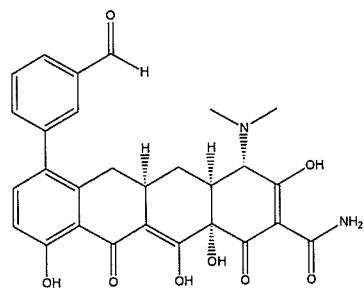
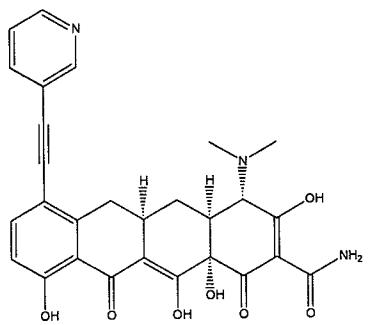


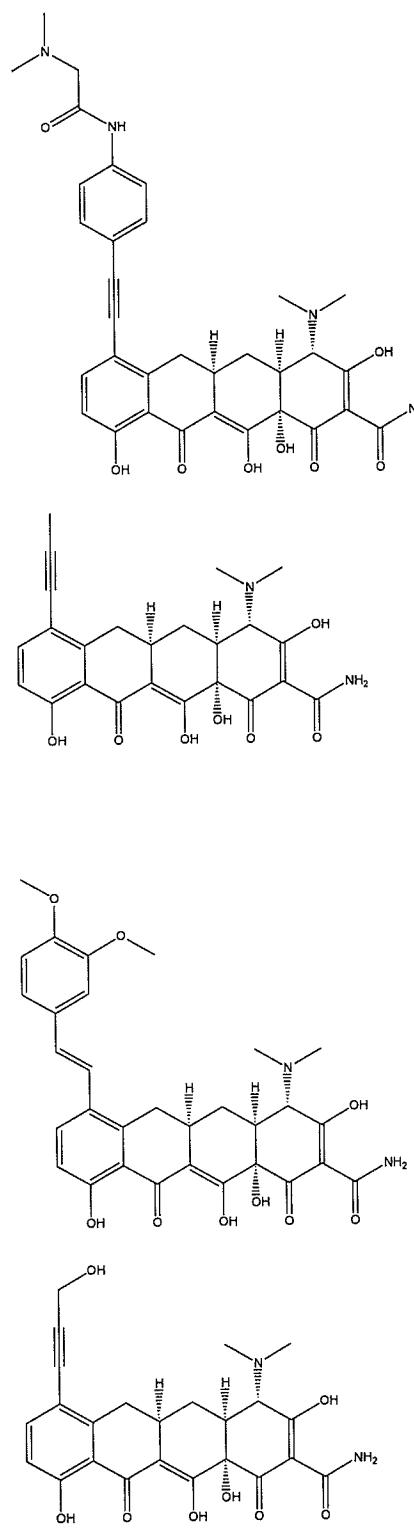
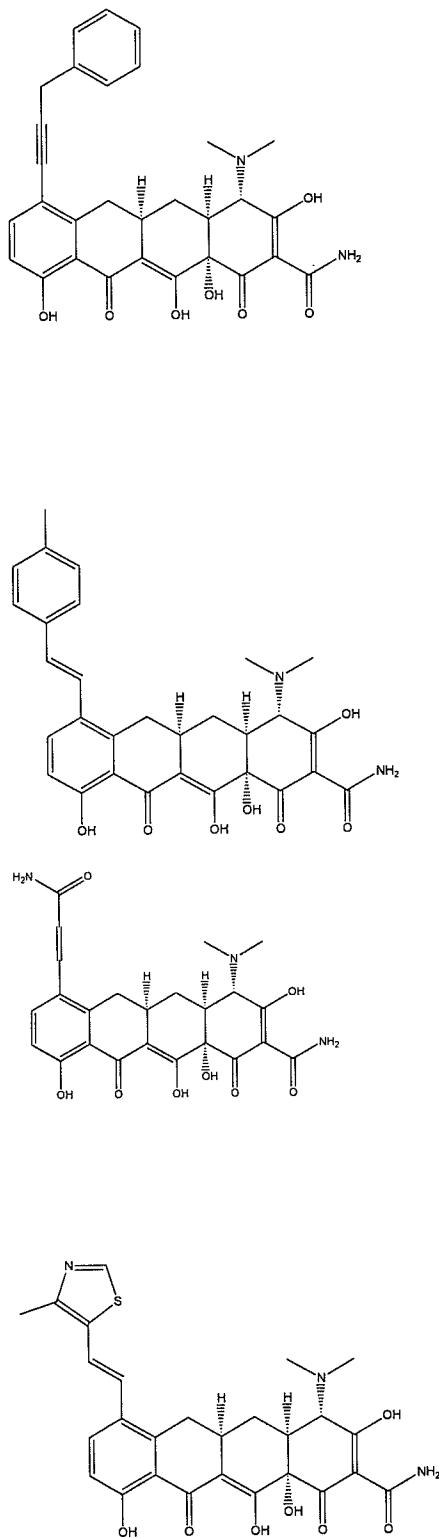


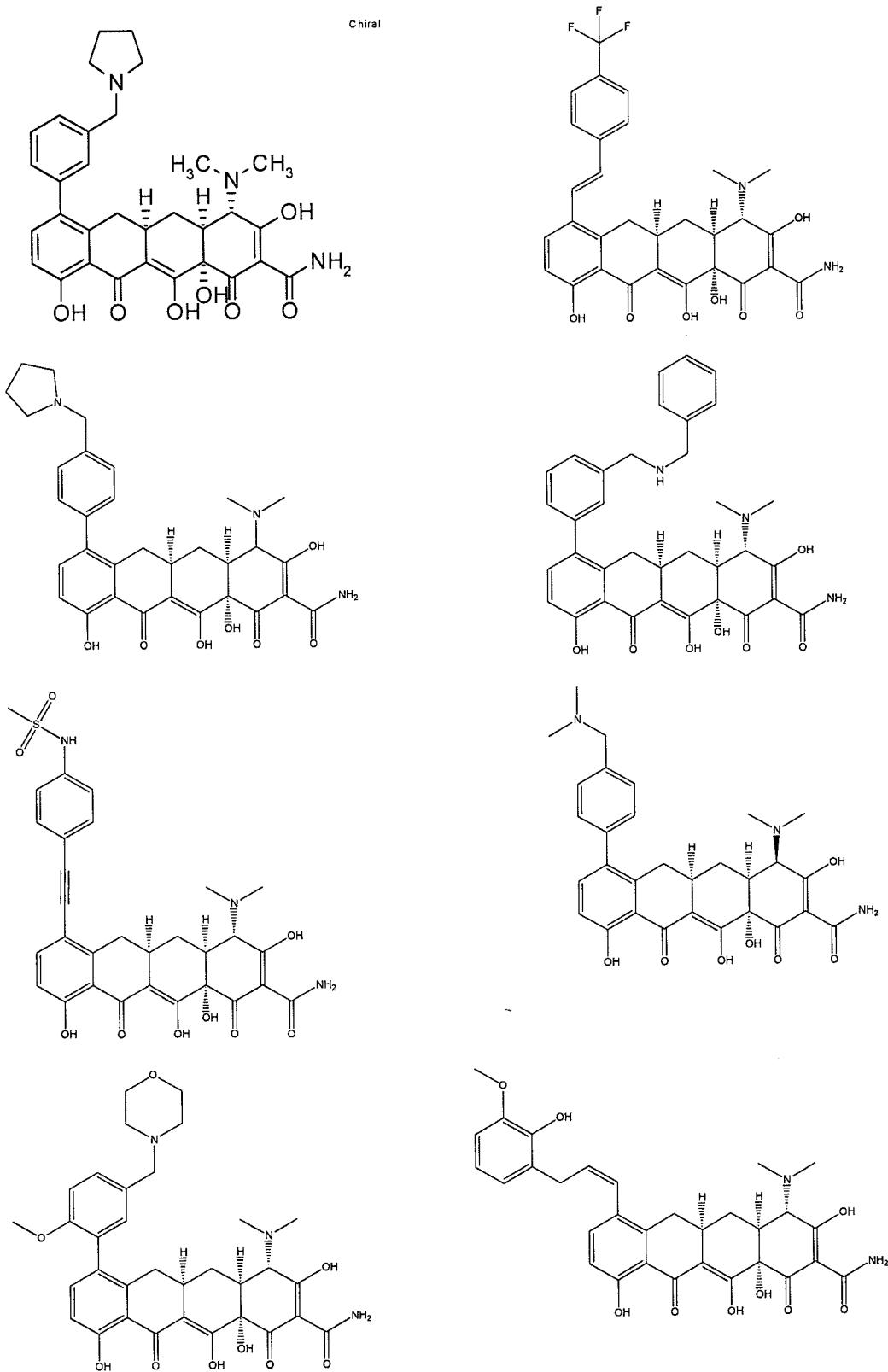


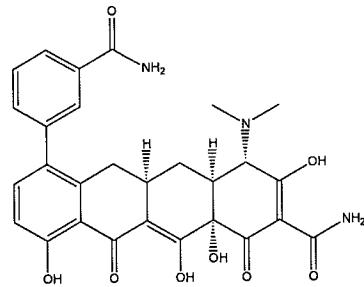
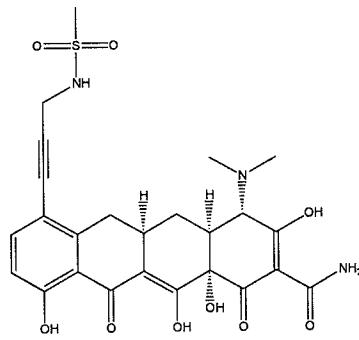
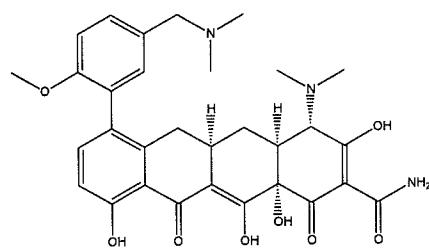
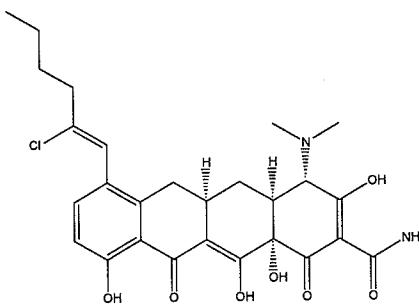
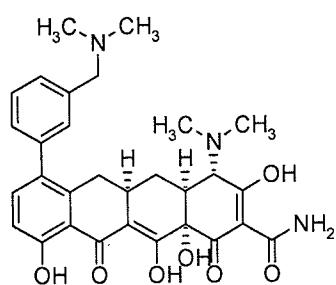
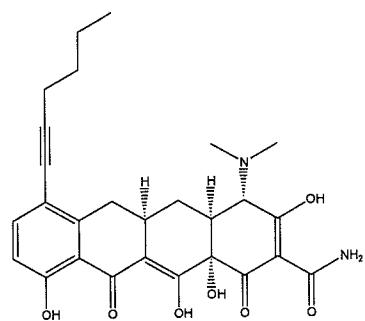
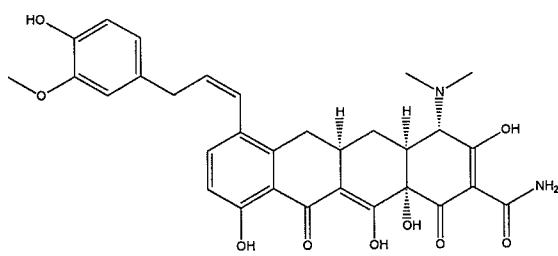


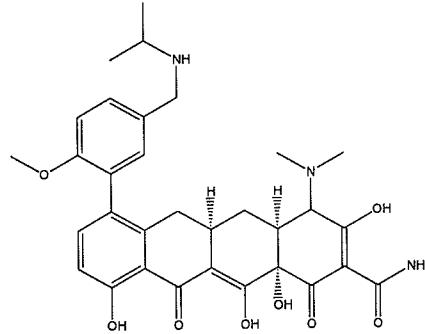
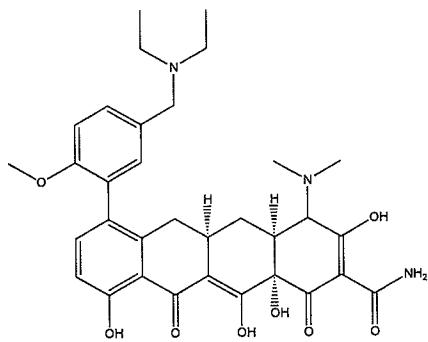
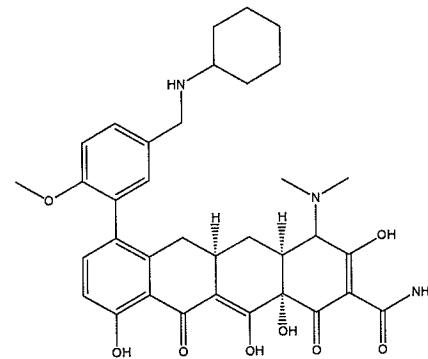
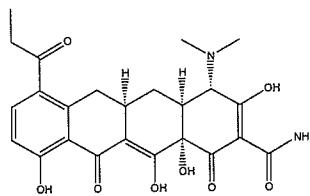
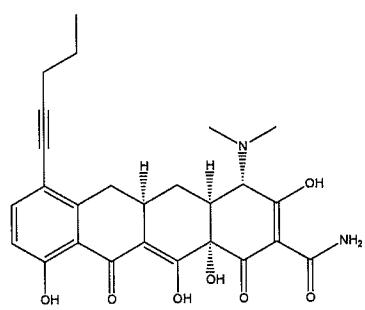
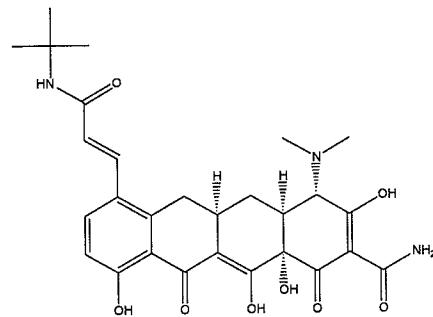
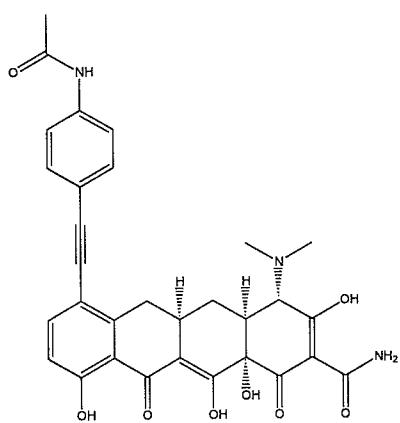


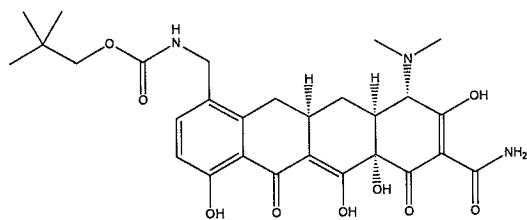
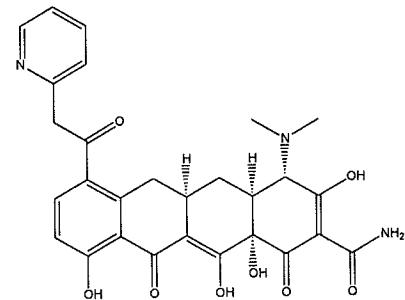
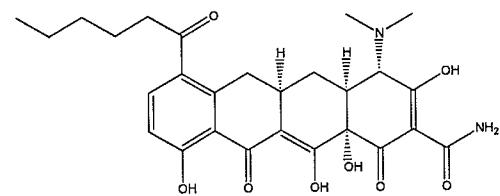
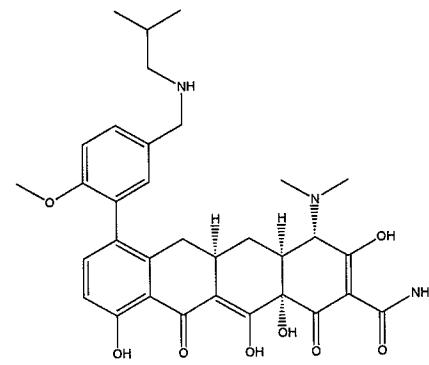
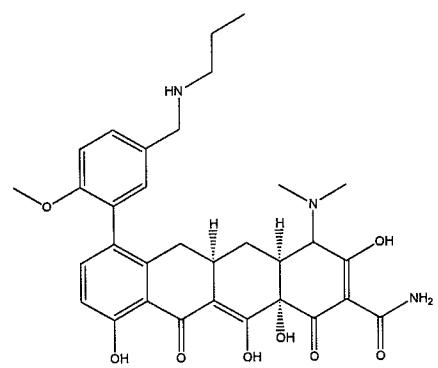
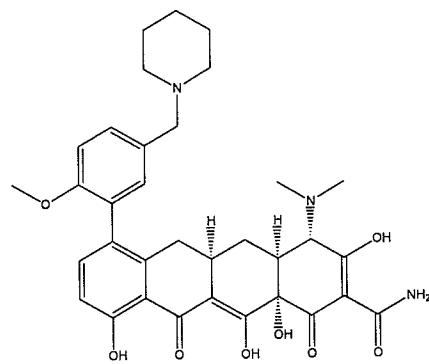
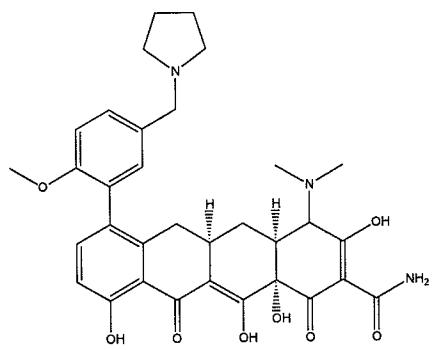


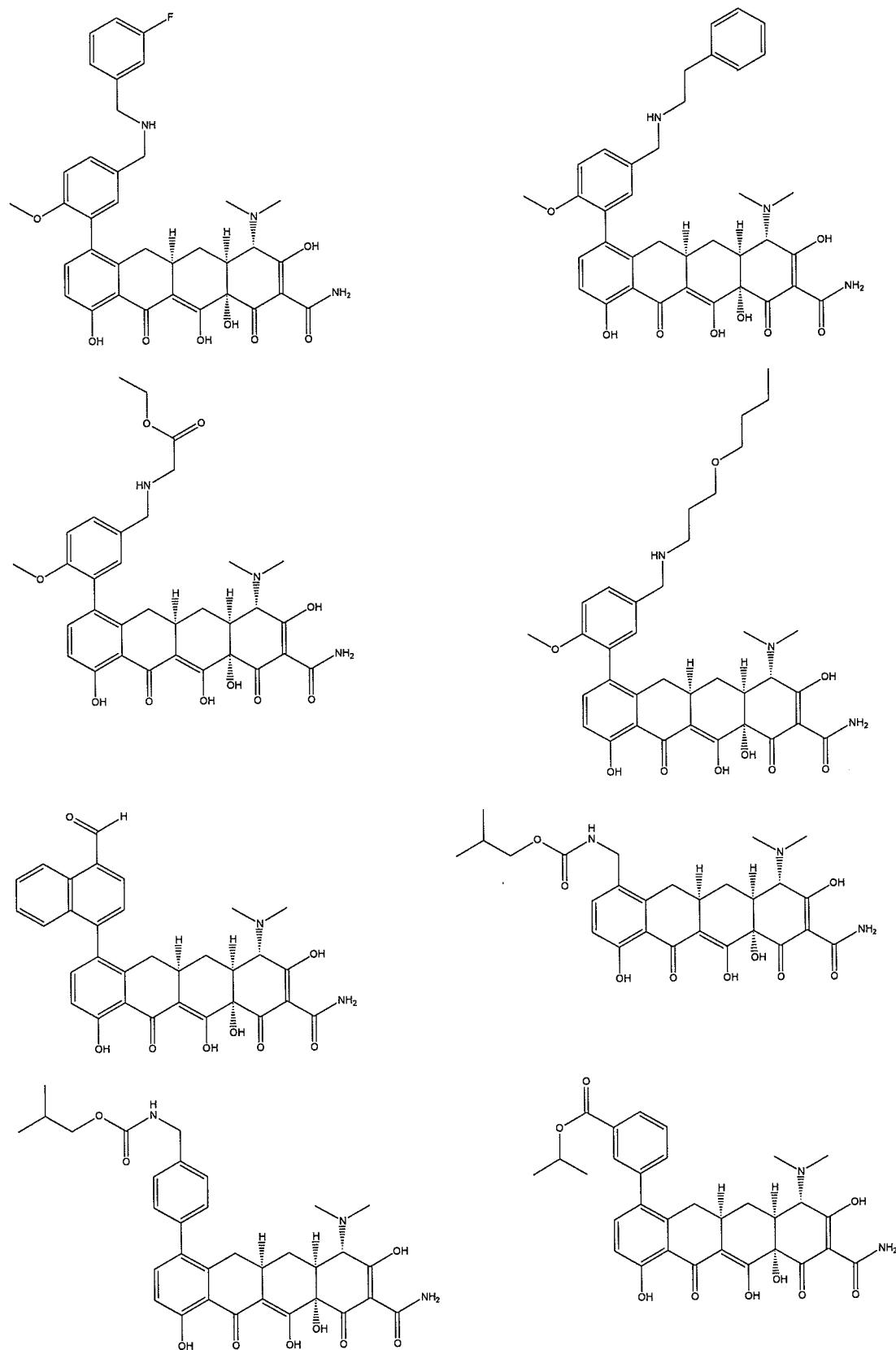


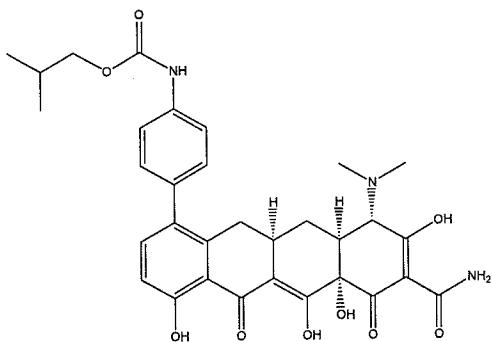
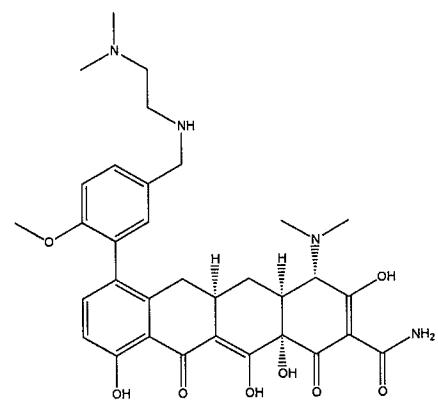
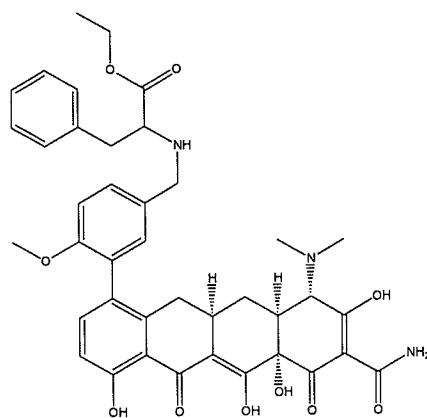
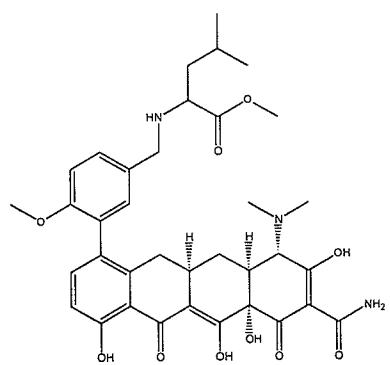
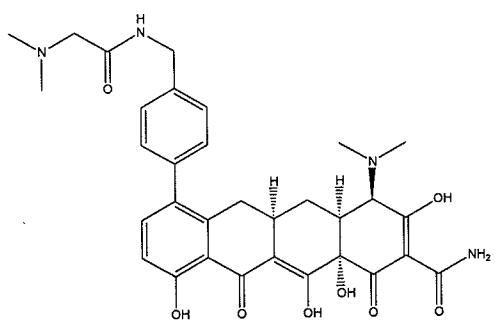
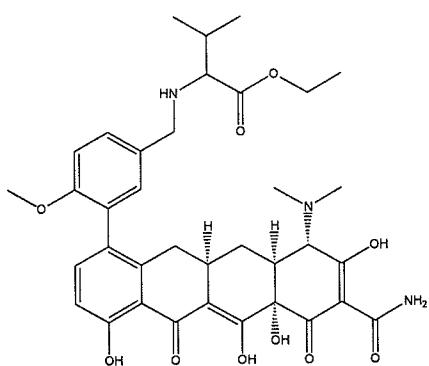


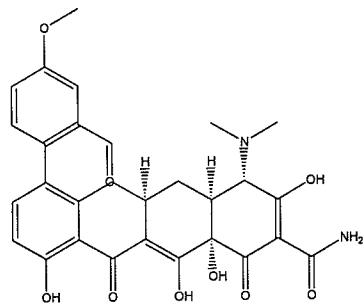
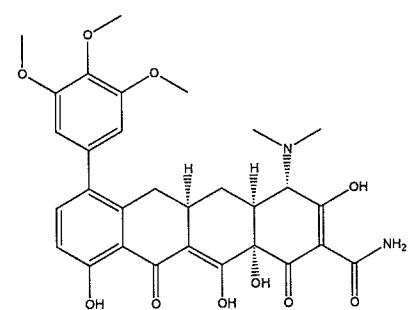
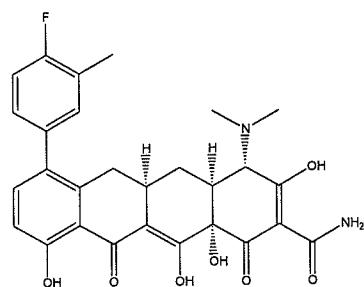
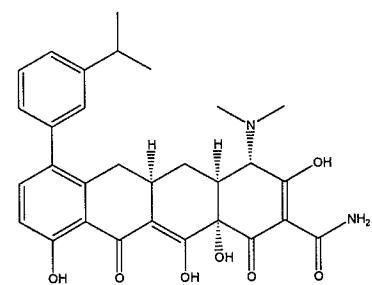
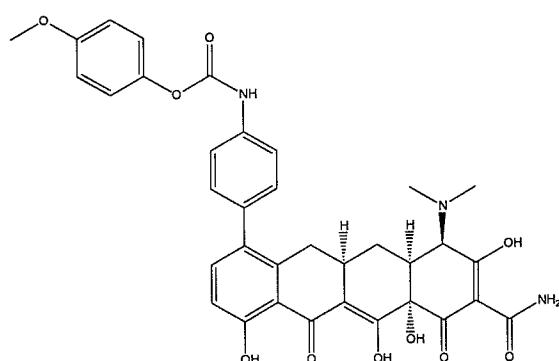
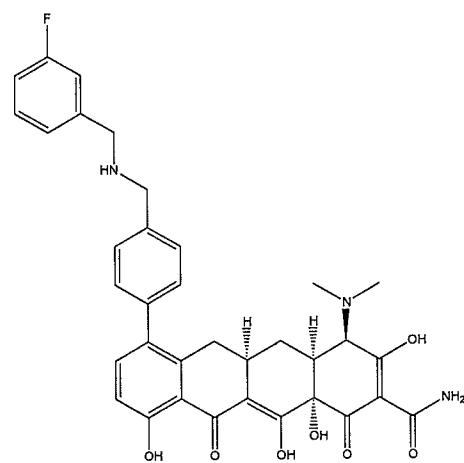
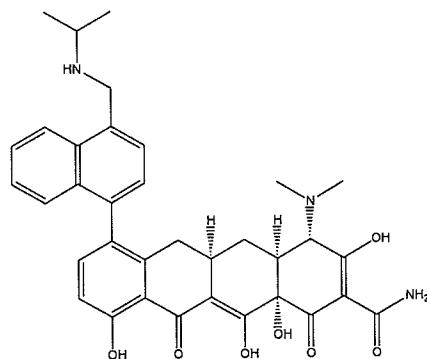
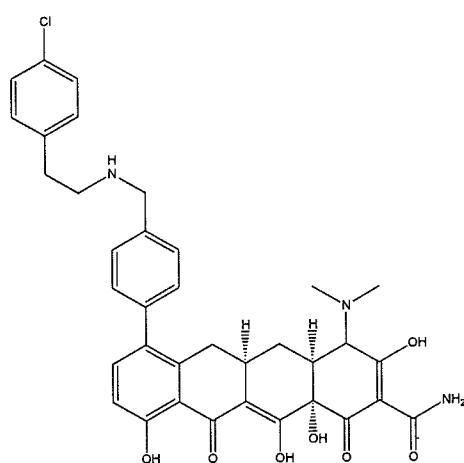


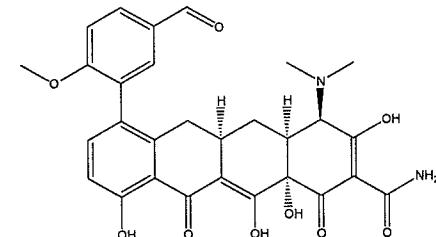
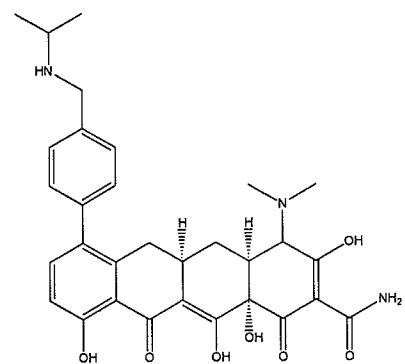
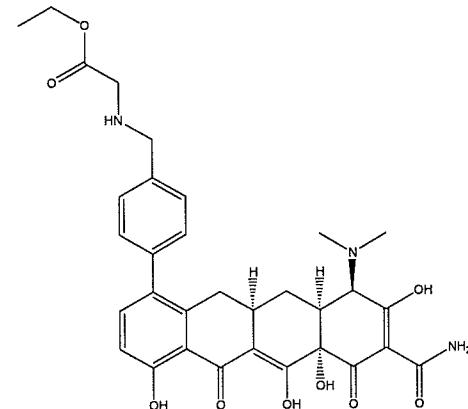
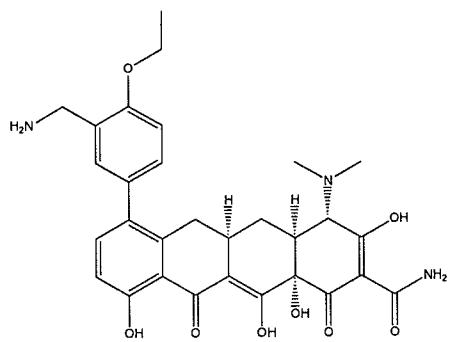
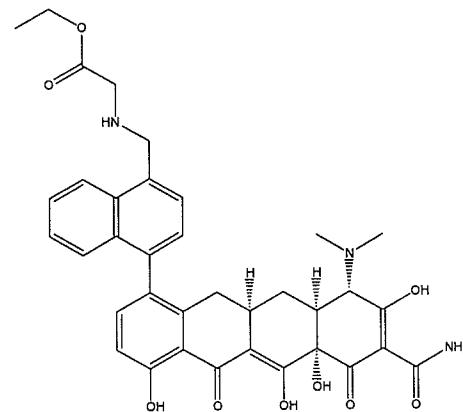
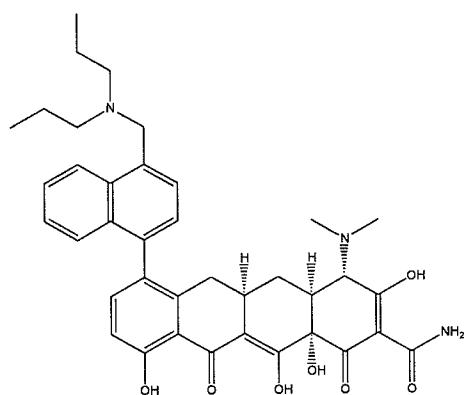
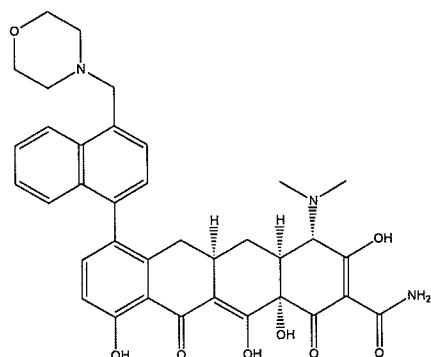
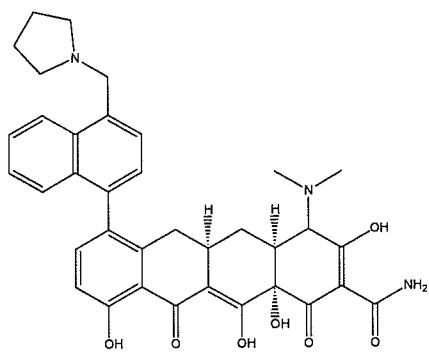


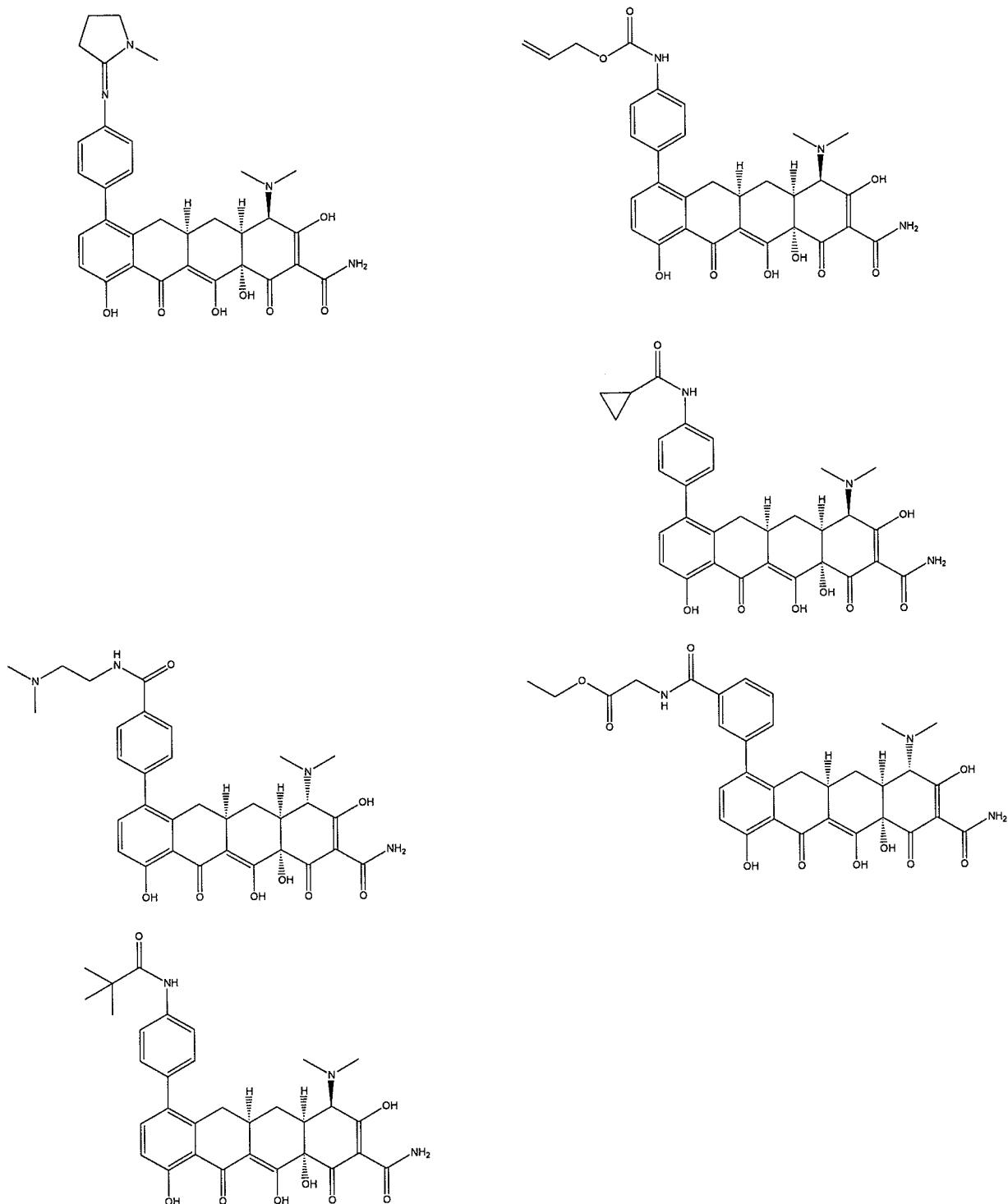


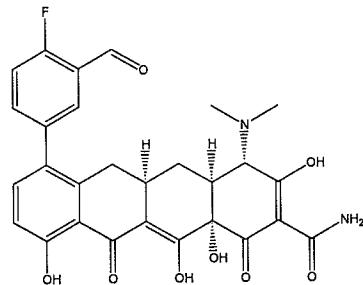
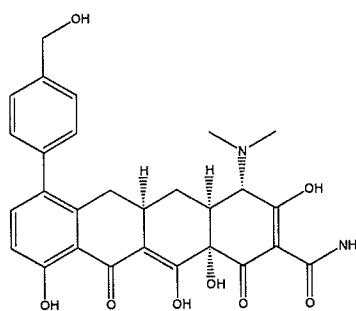
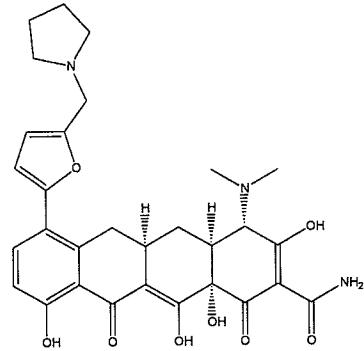
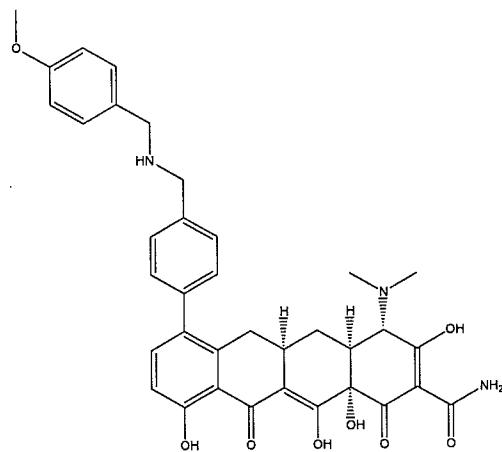
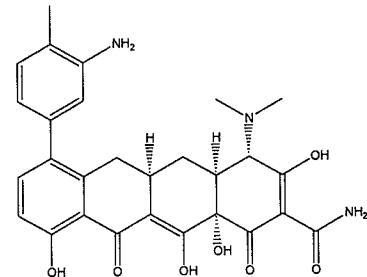
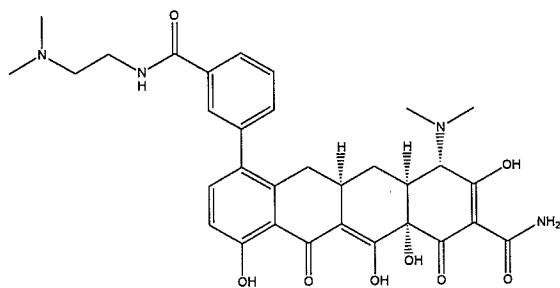
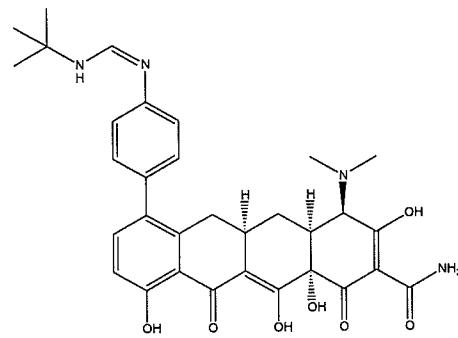
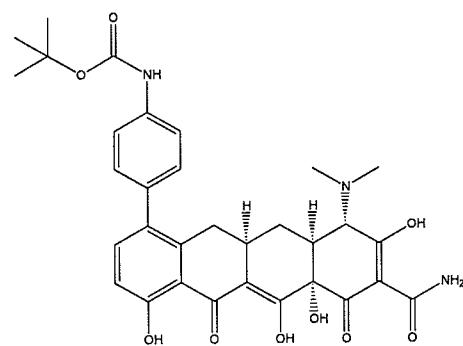


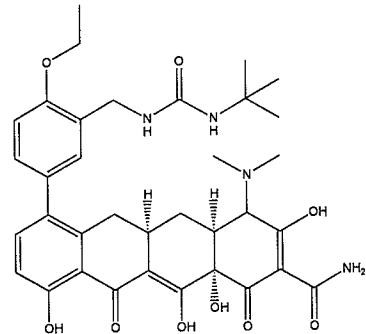
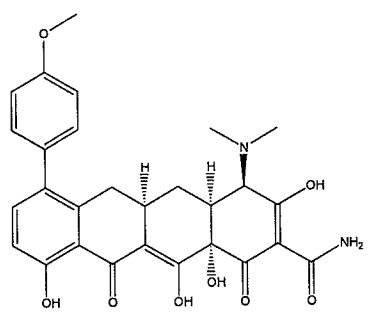
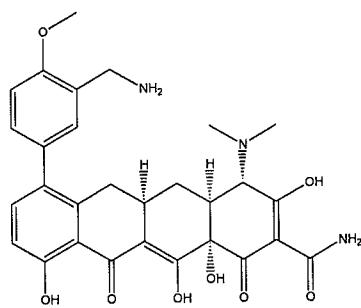
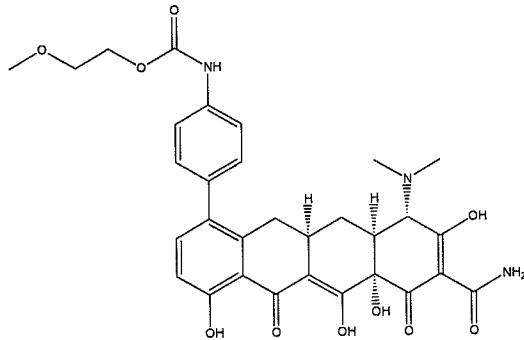
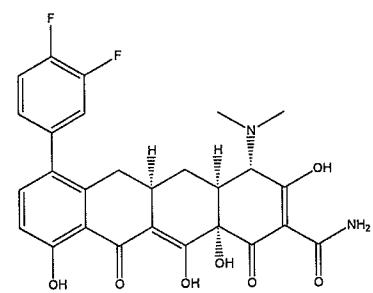
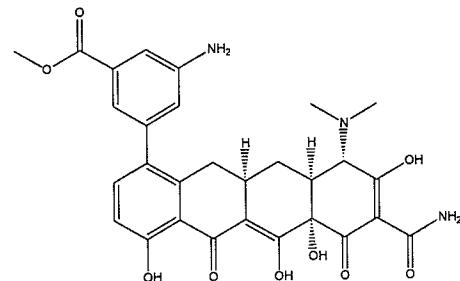
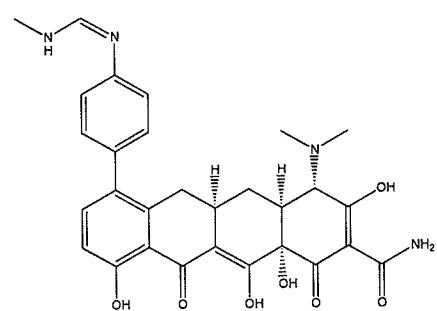
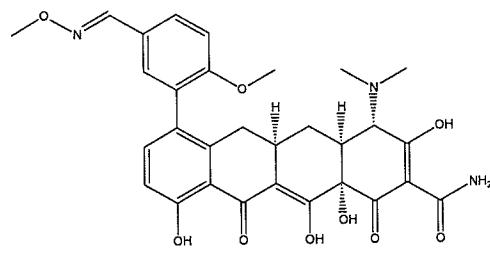
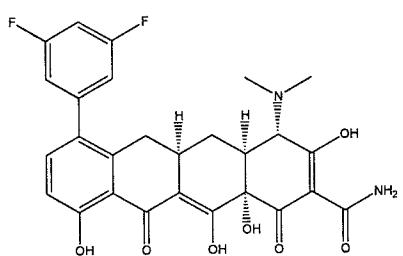


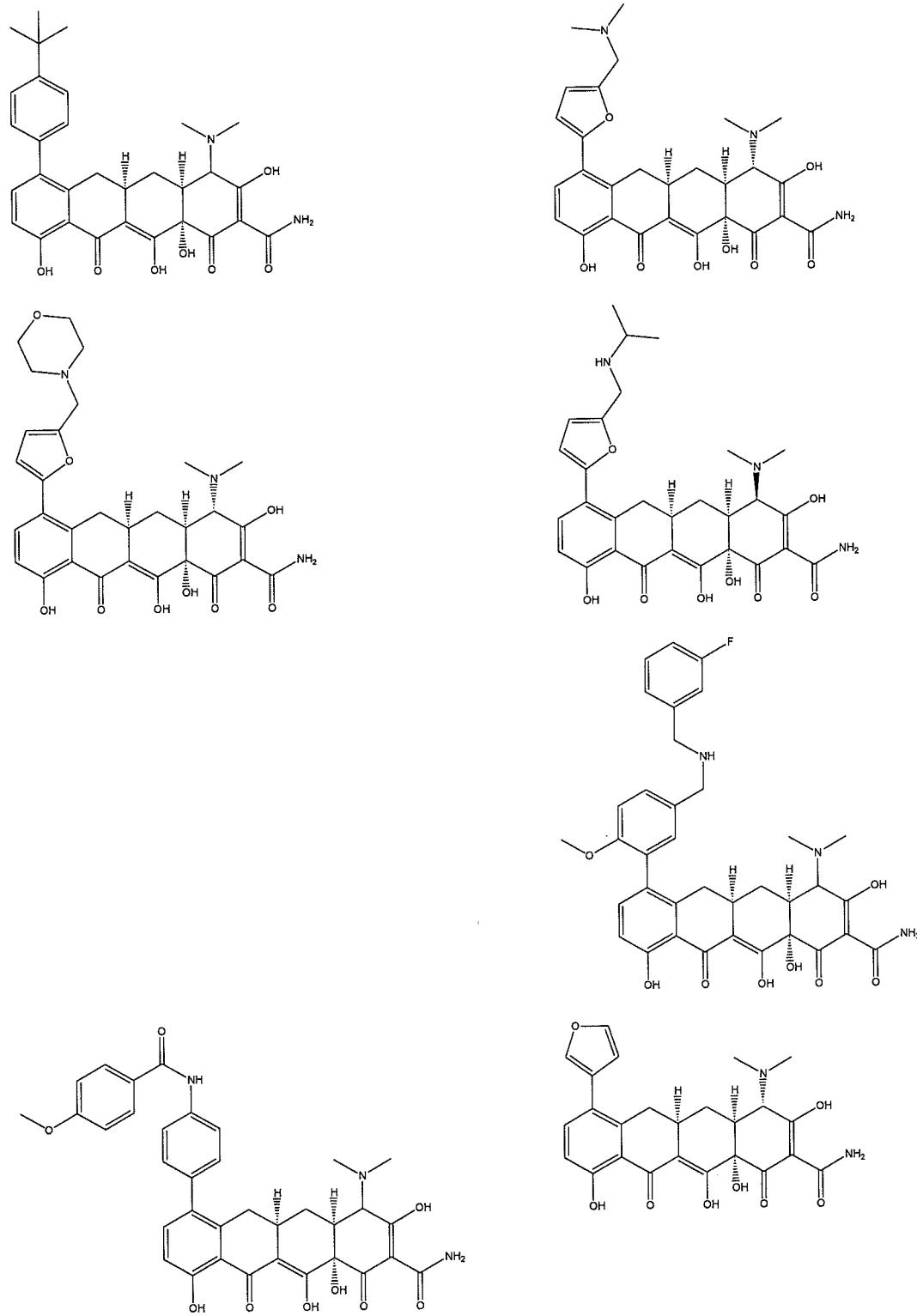


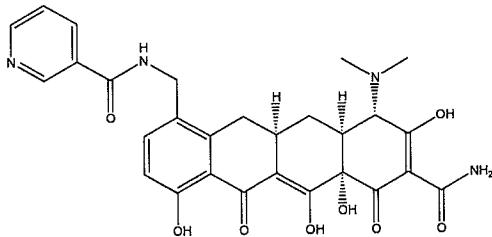
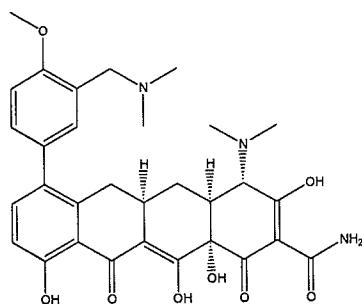
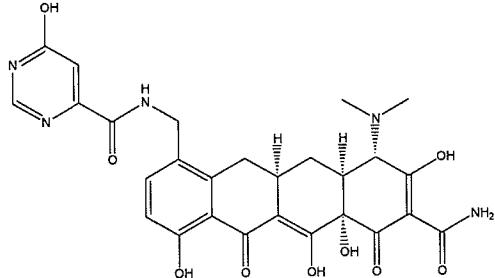
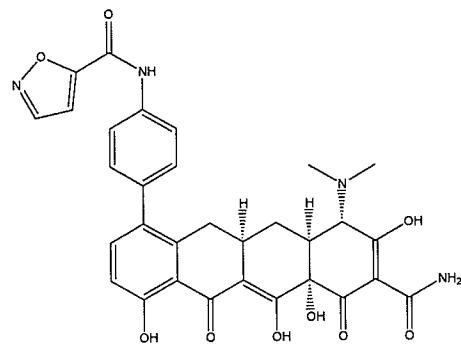
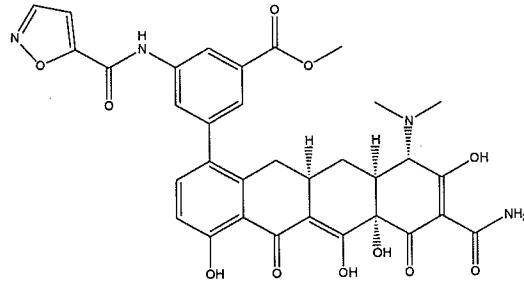
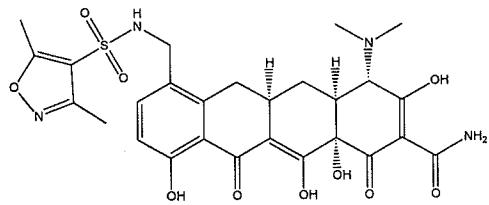
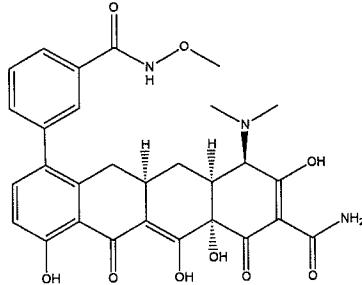
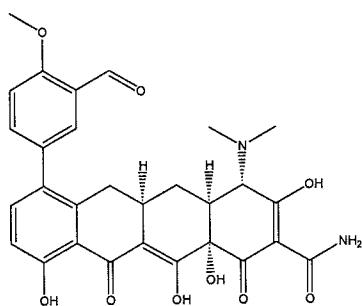
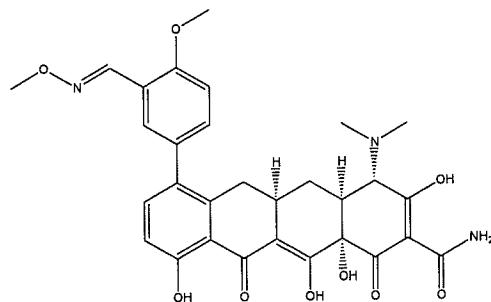
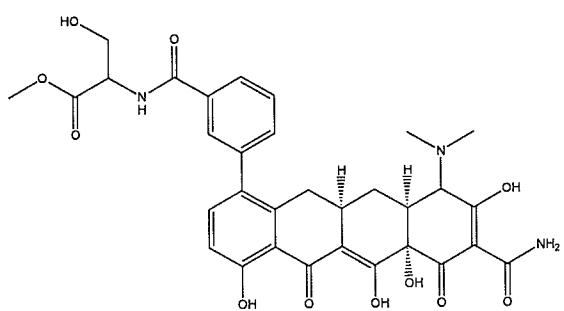


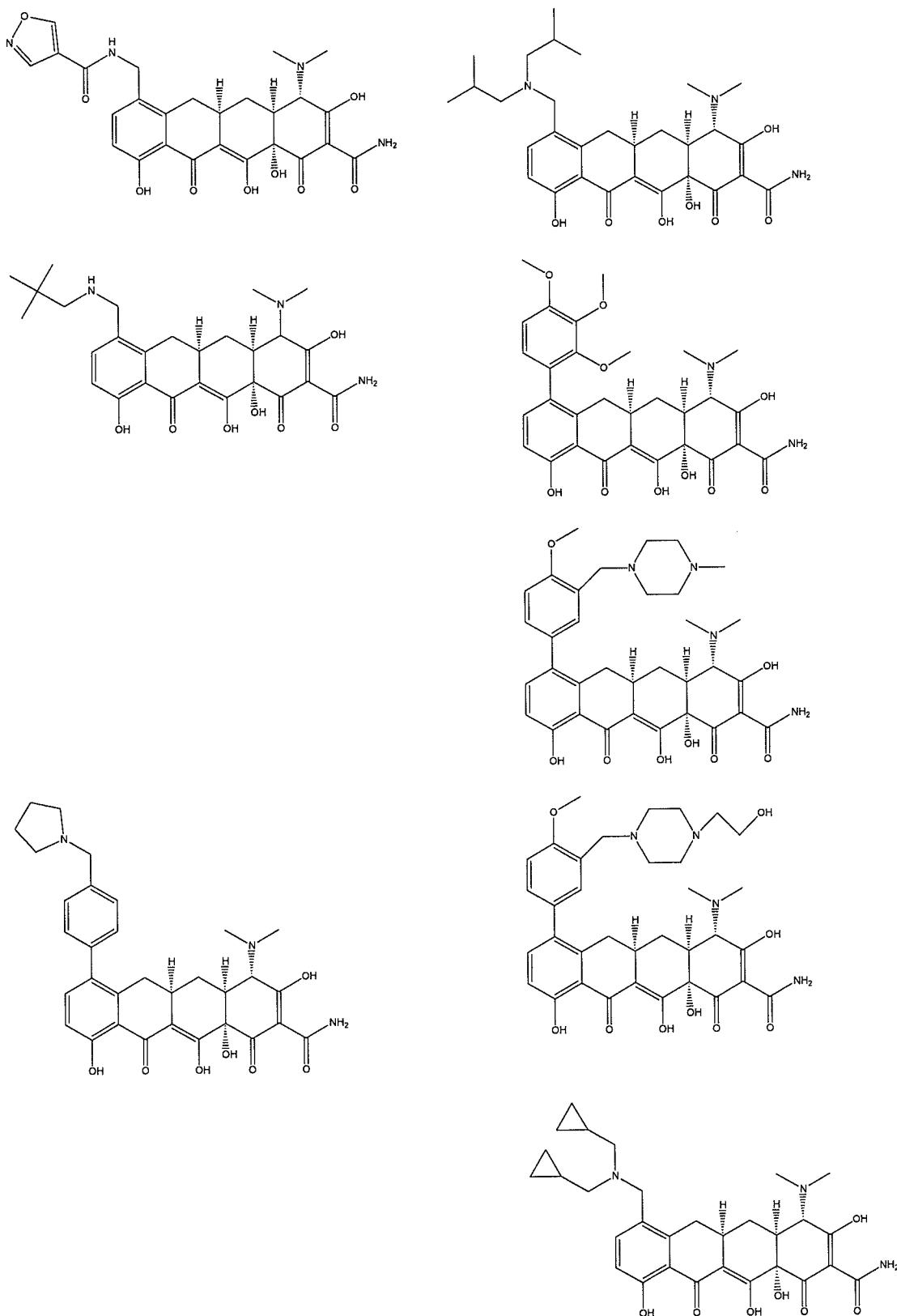


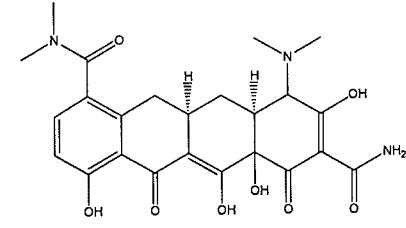
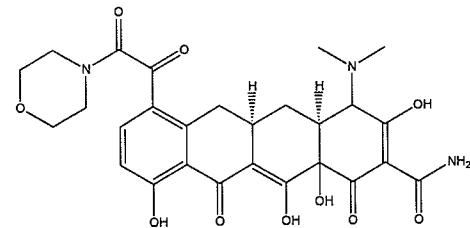
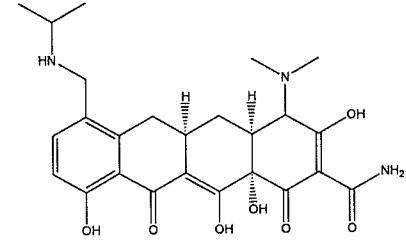
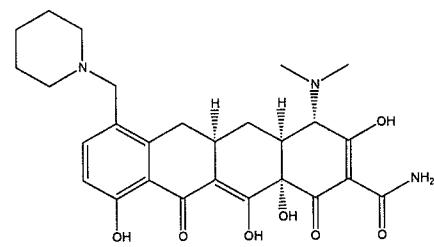
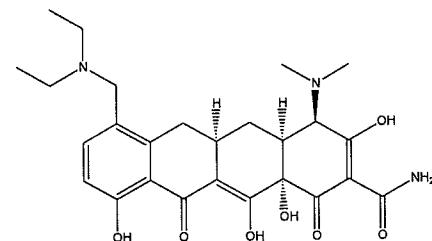
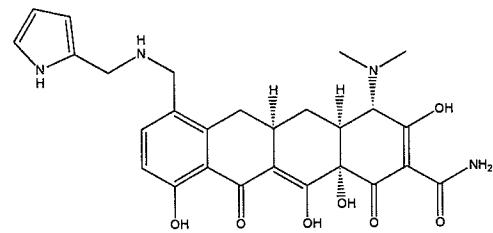
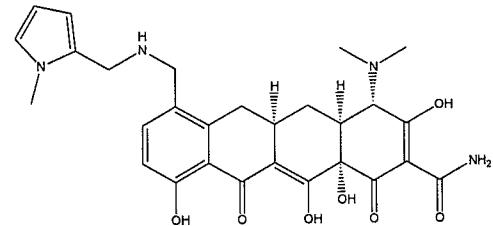
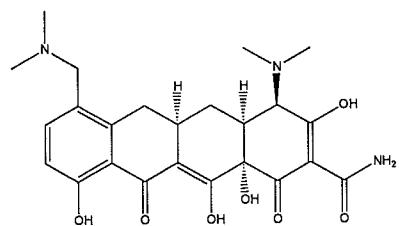
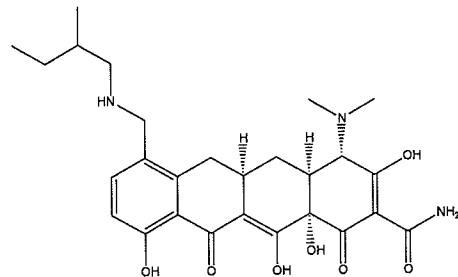
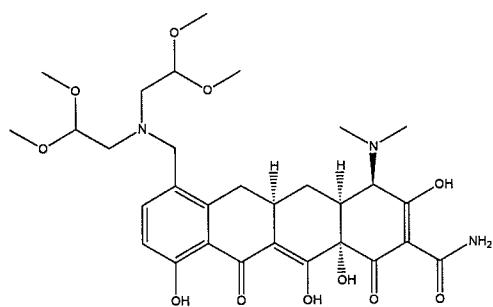


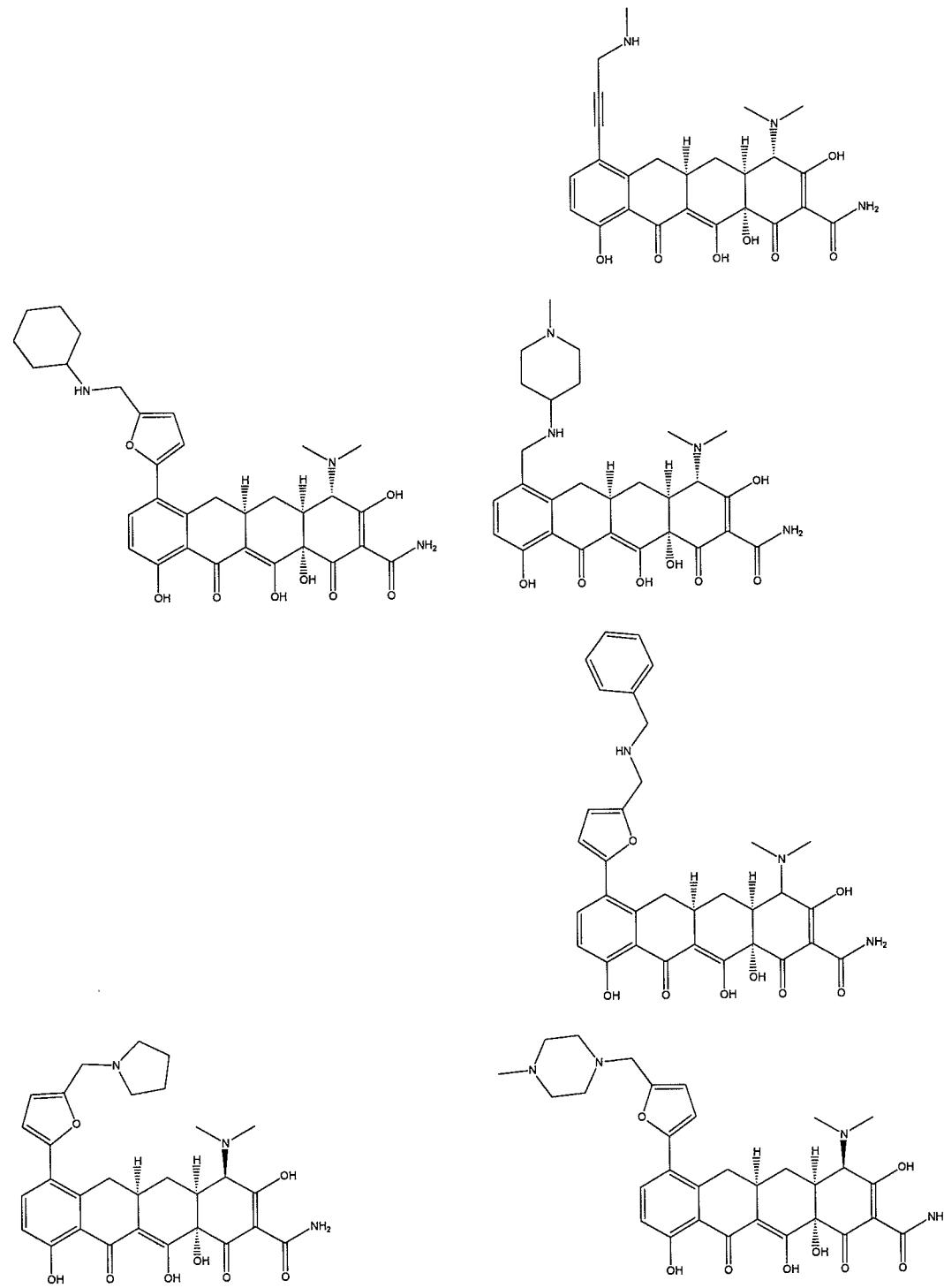


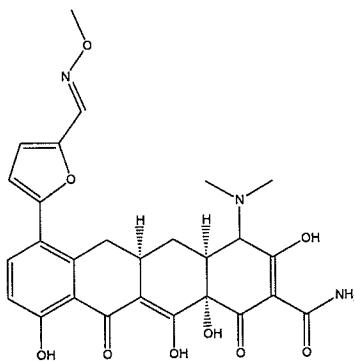
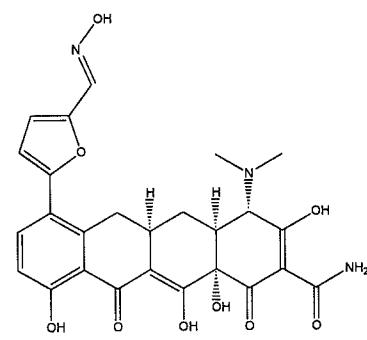
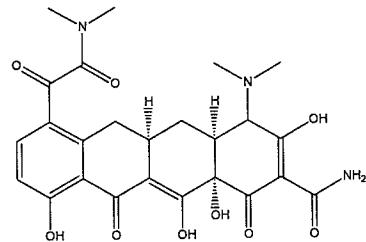
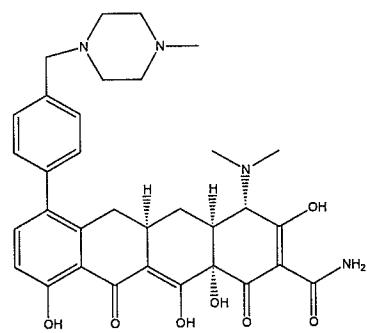
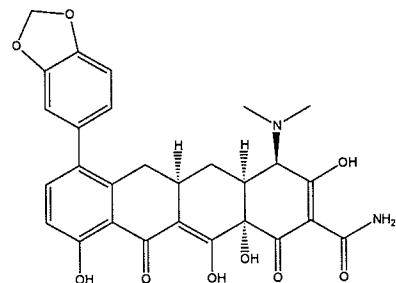
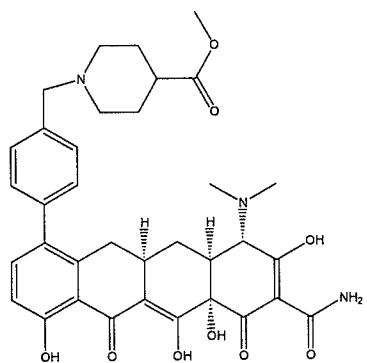
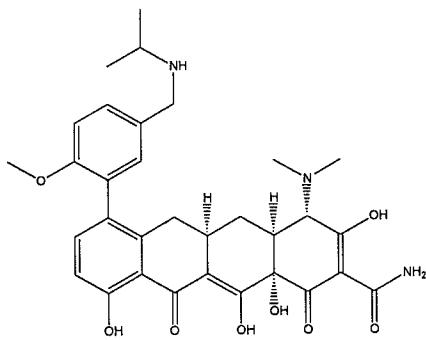


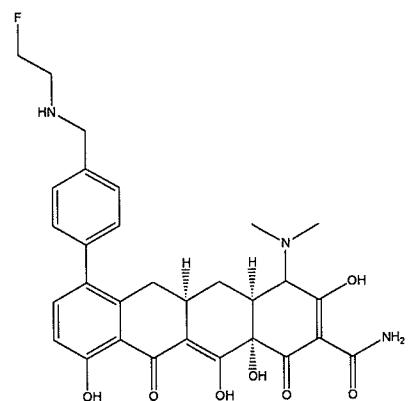
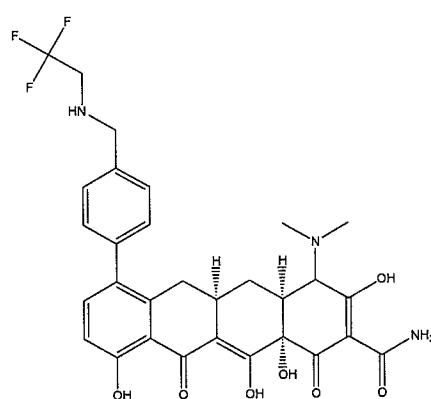
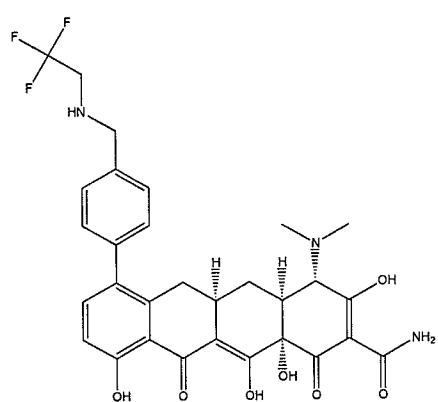
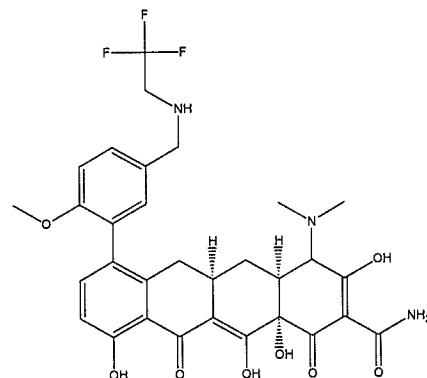
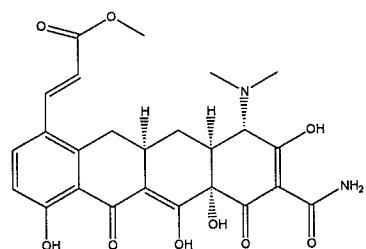
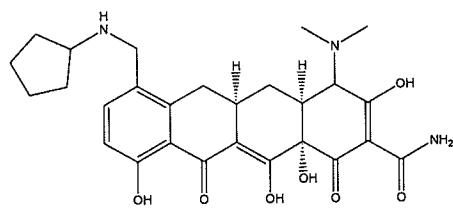
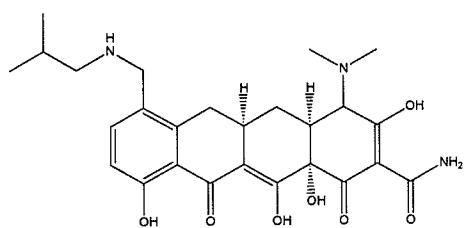


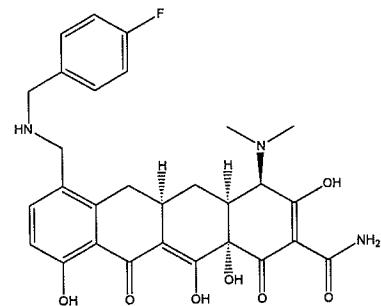
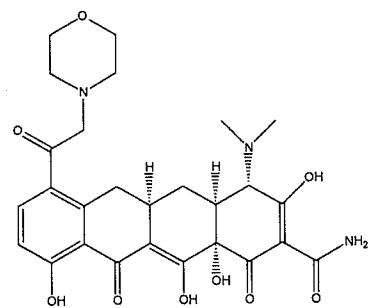
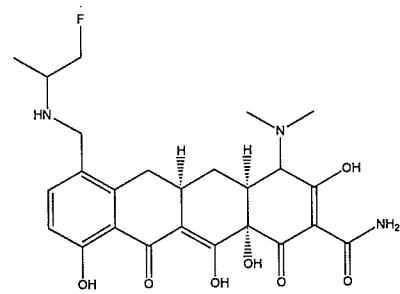
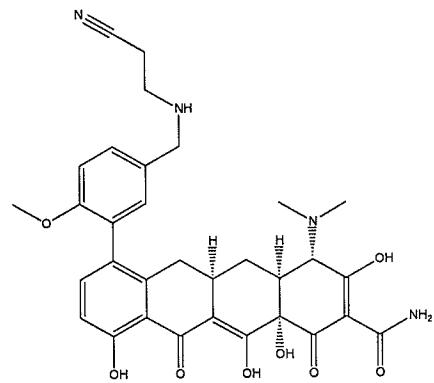
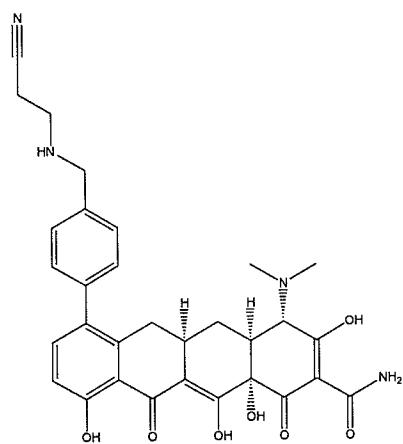
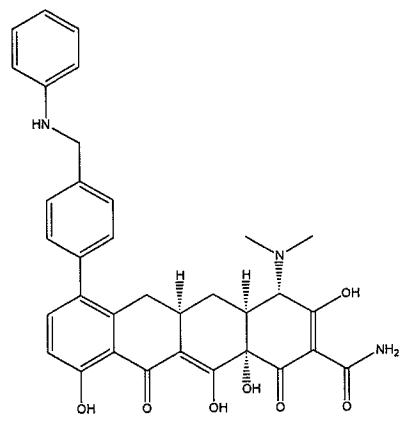


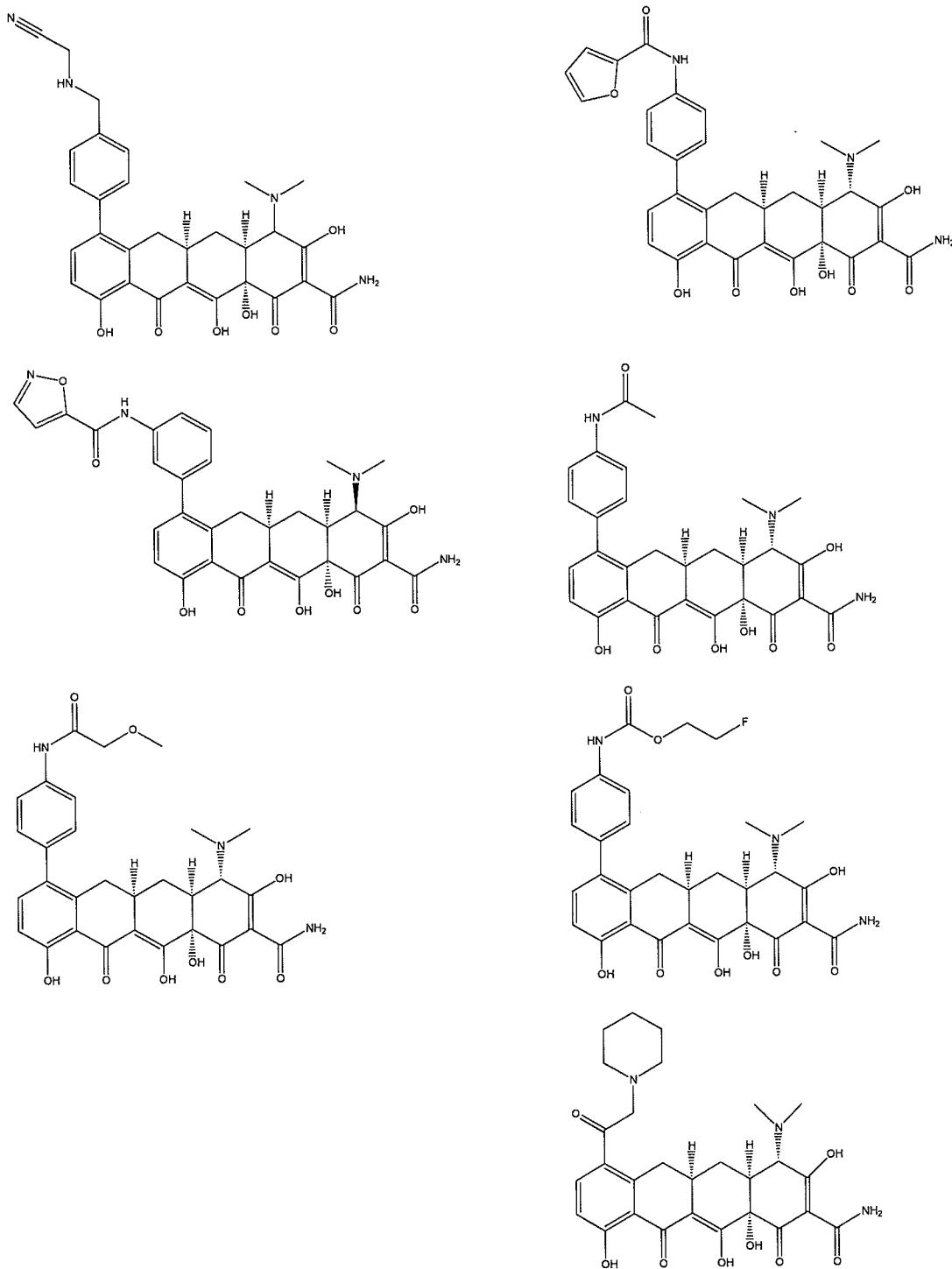






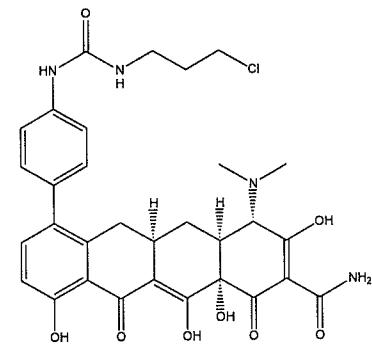
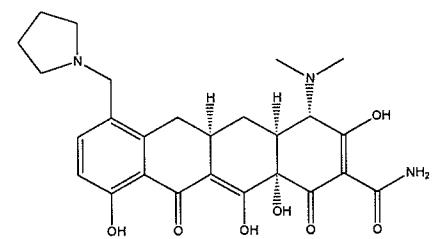
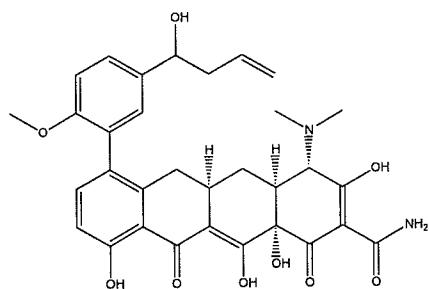
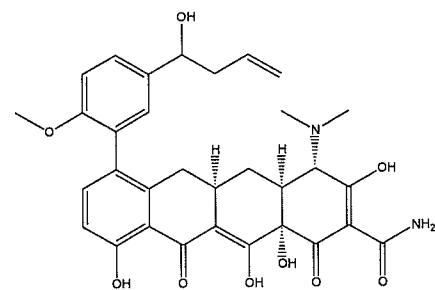
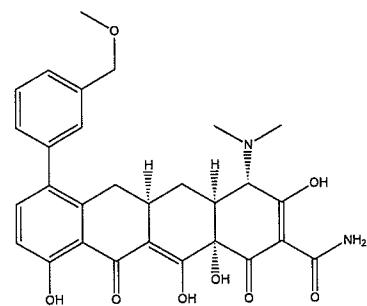
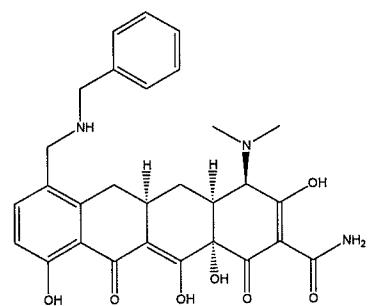
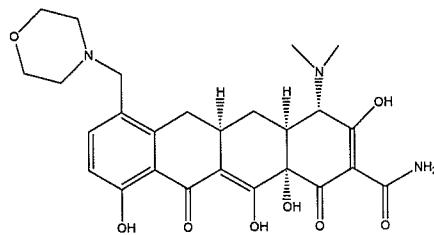
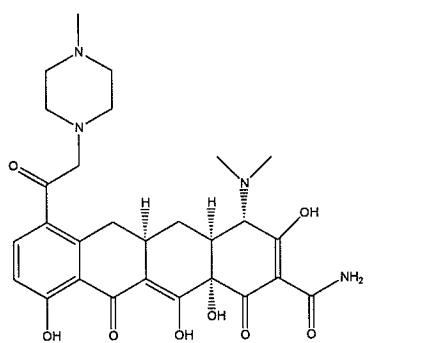


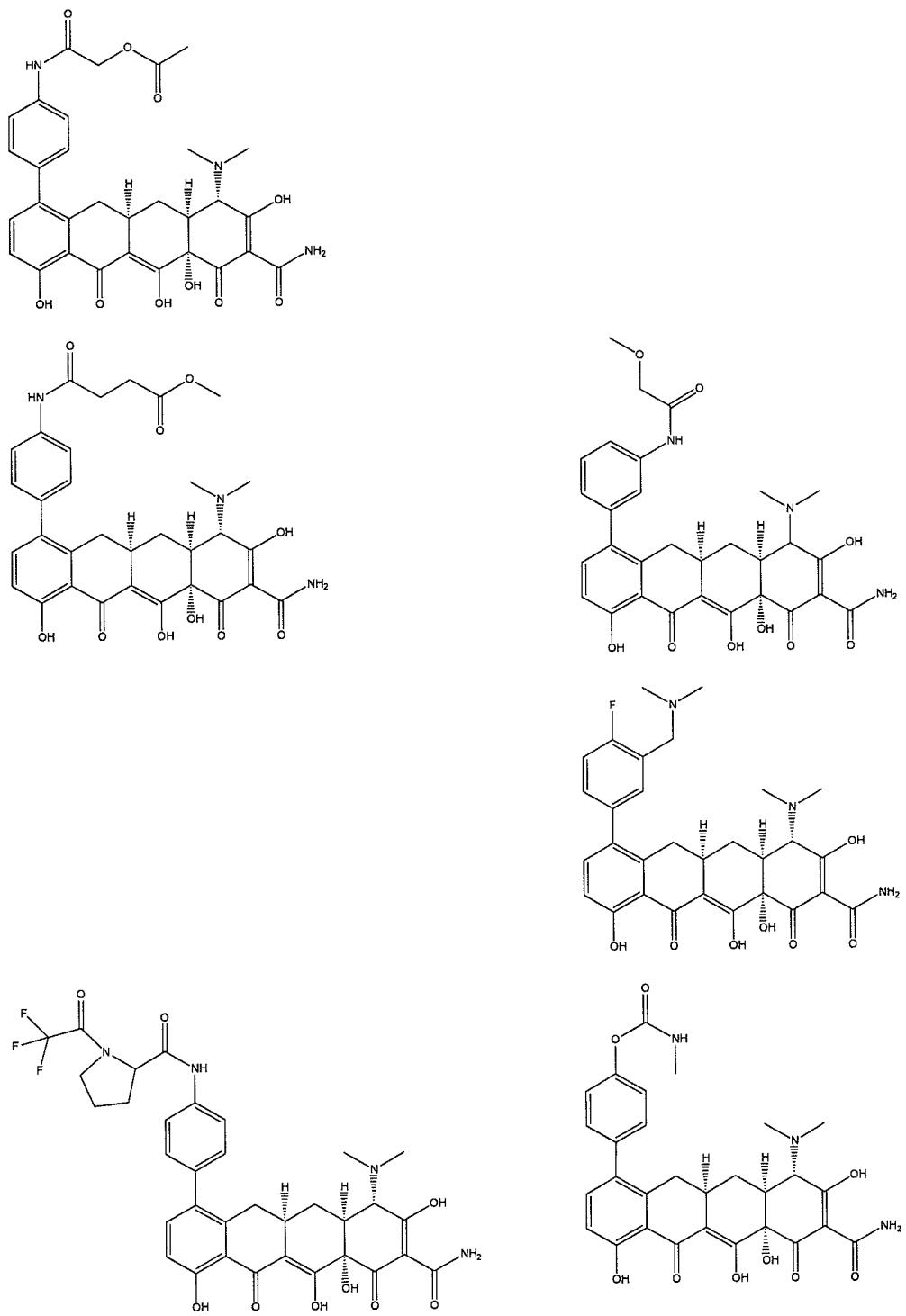


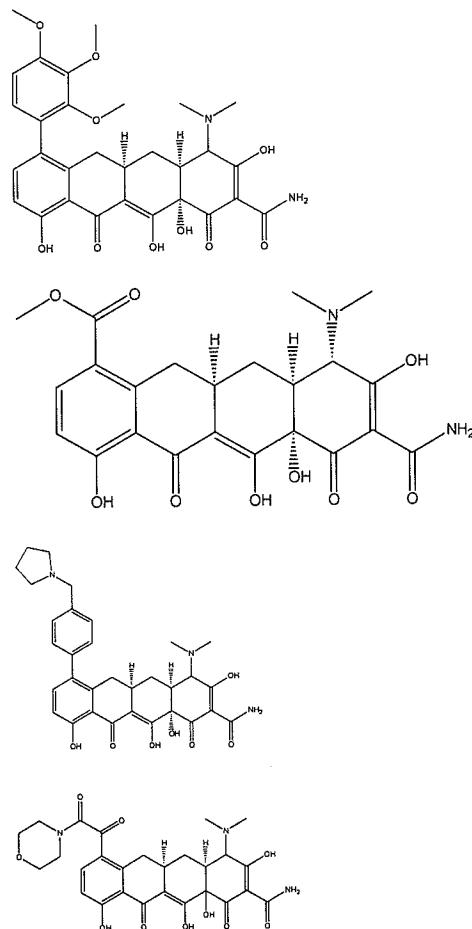
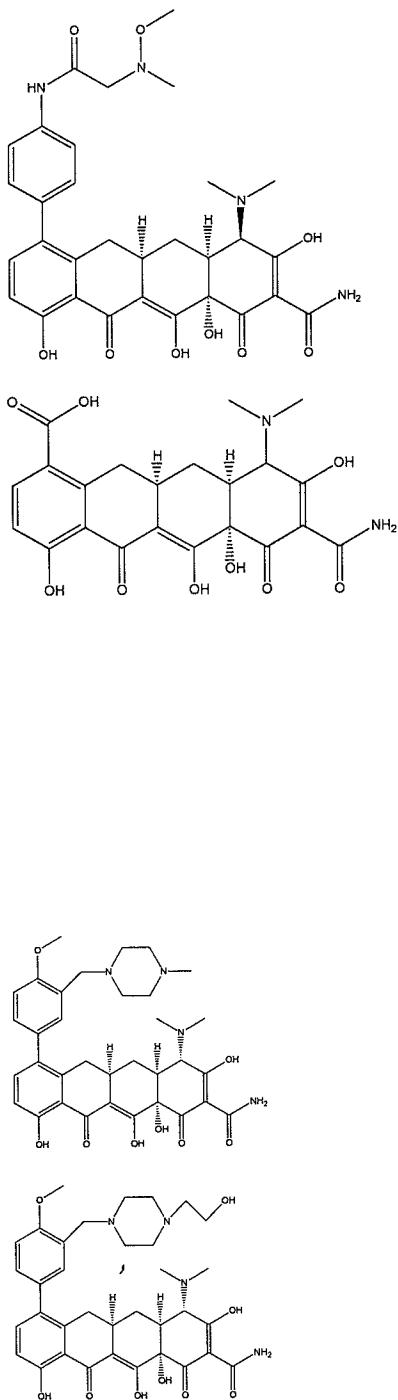


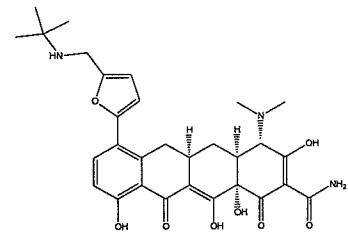
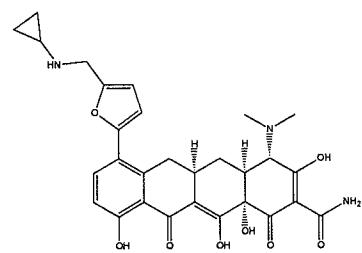
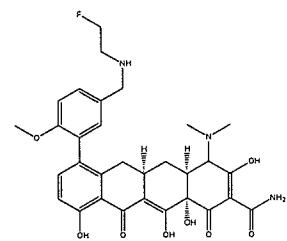
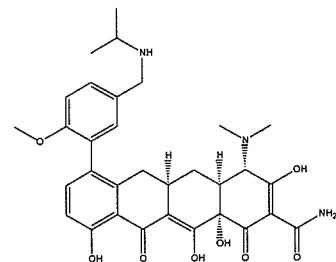
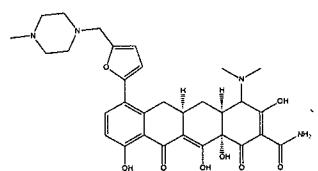
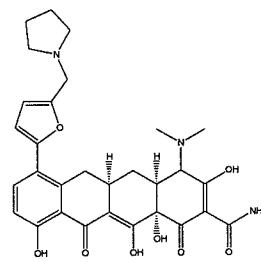
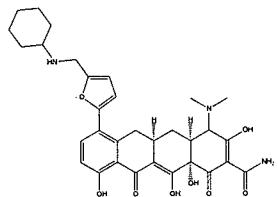
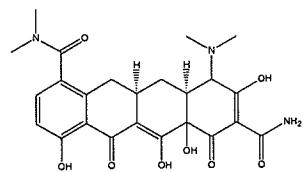
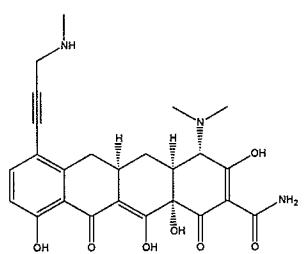
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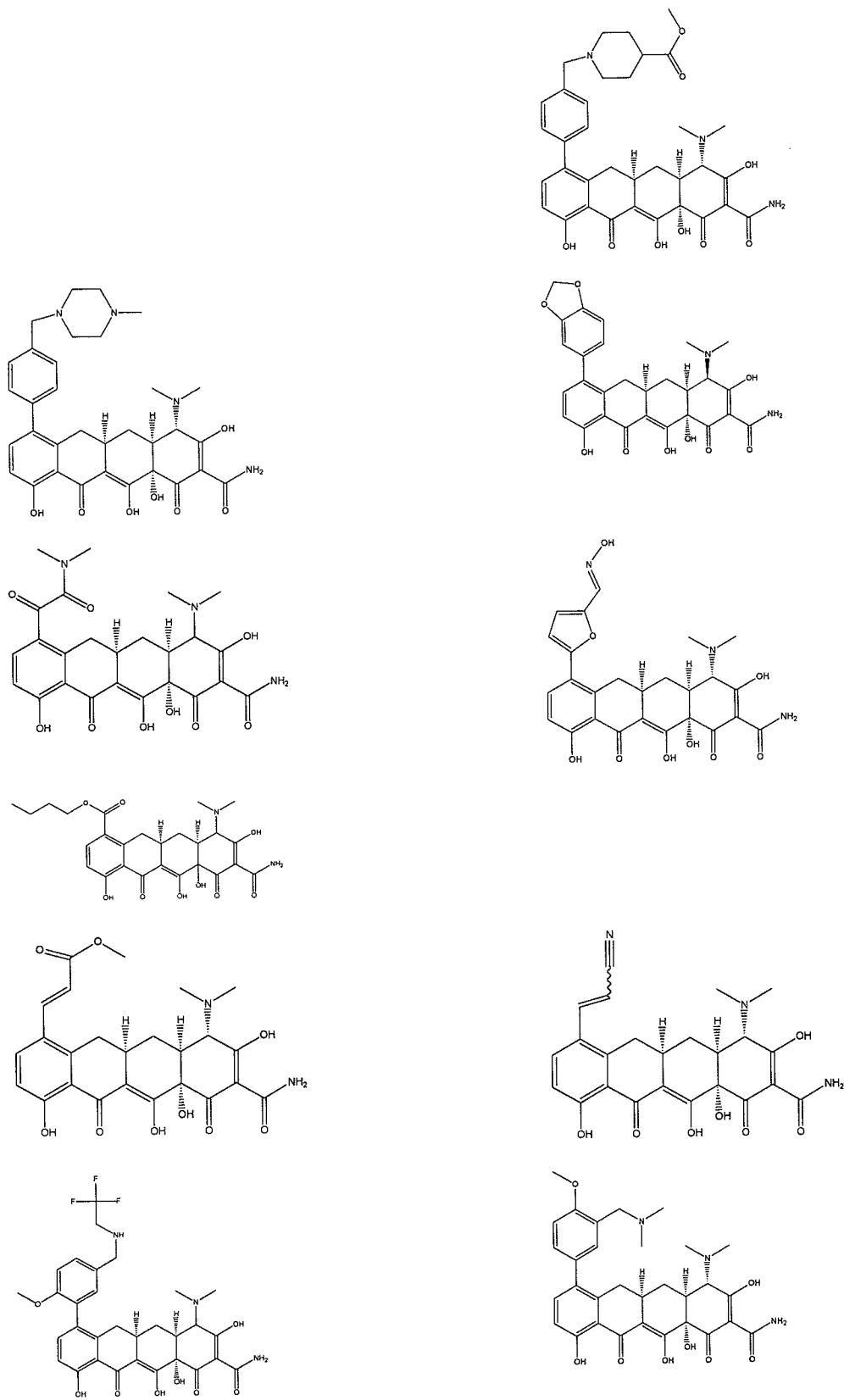
Docket No.: 16534-539001US

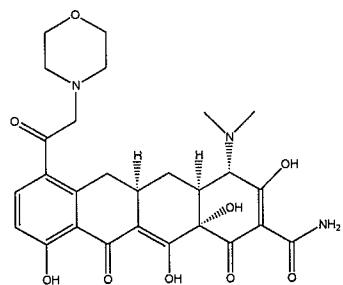
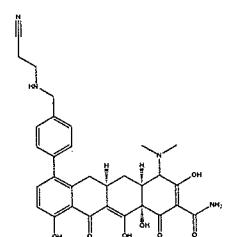
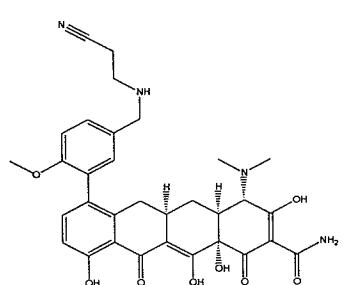
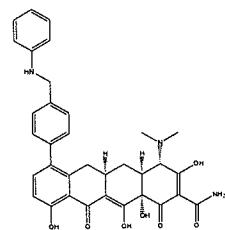
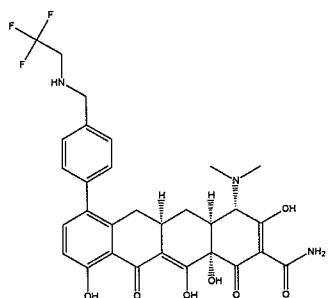
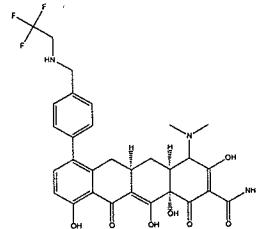
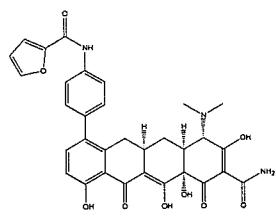


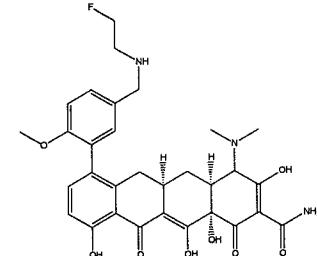
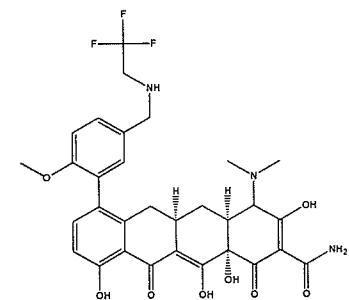
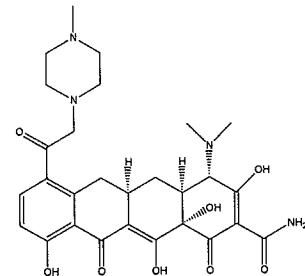
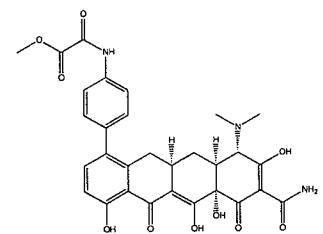
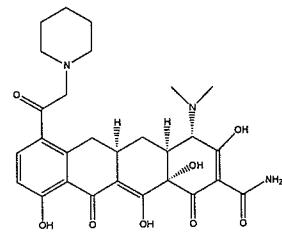
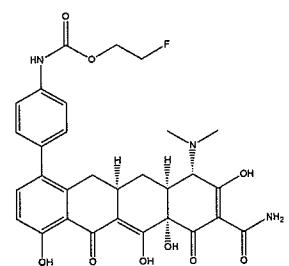
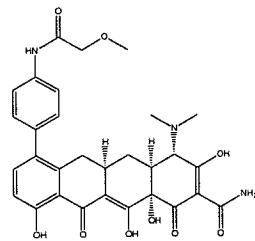
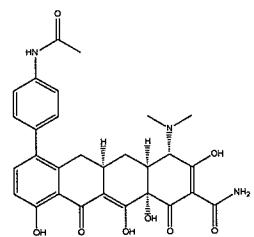
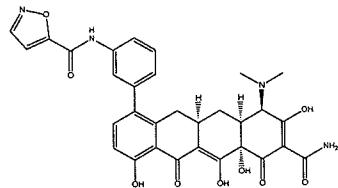
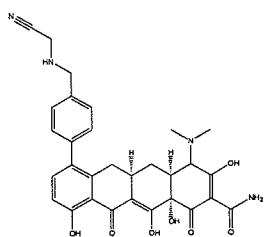


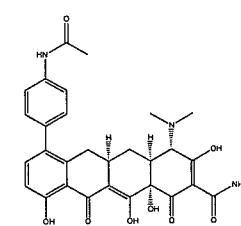
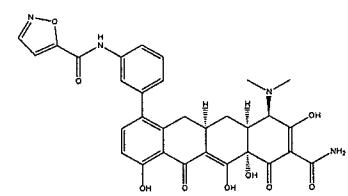
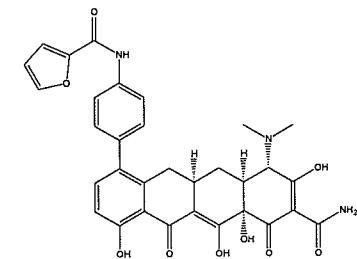
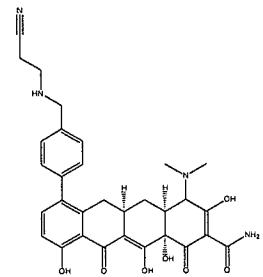
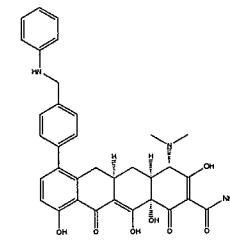
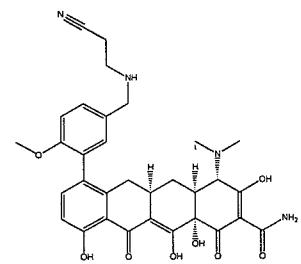
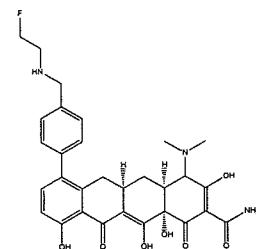
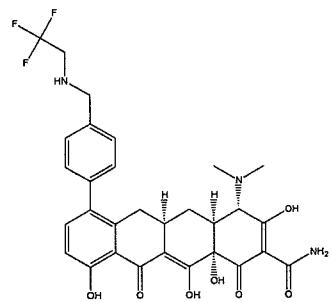
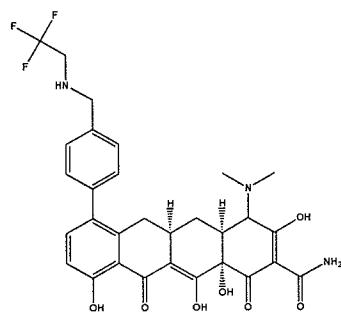


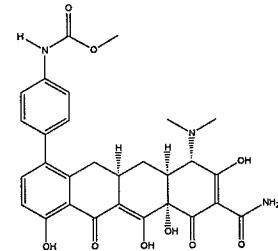
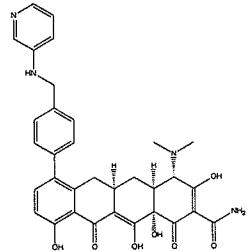
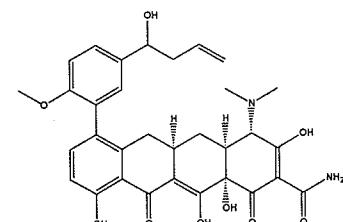
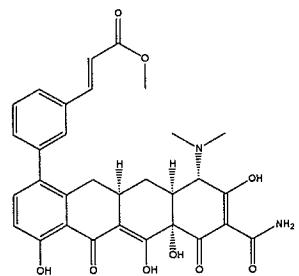
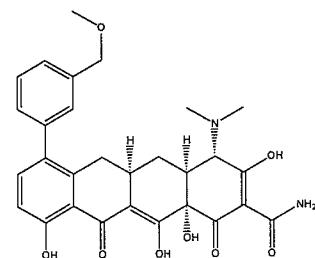
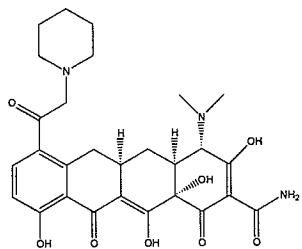
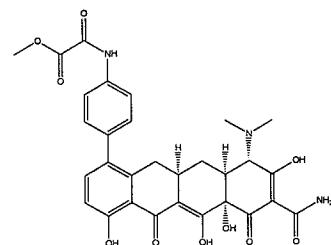
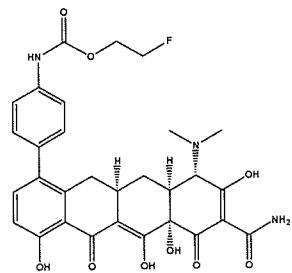
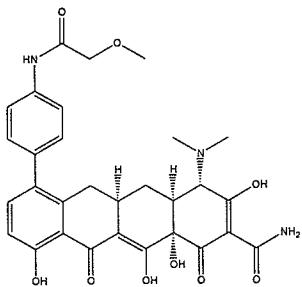


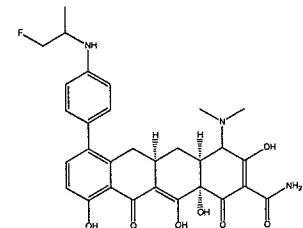
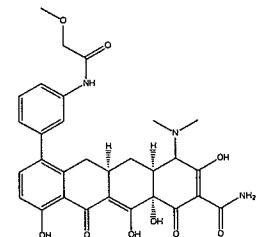
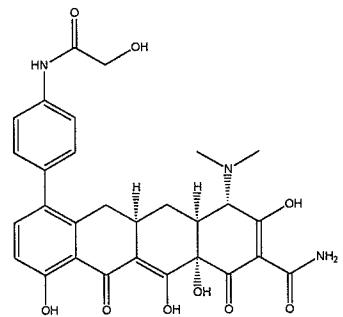
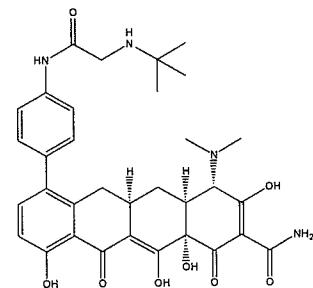
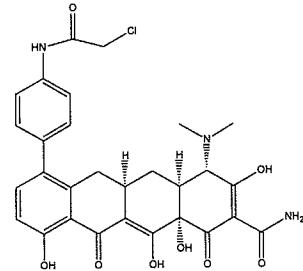
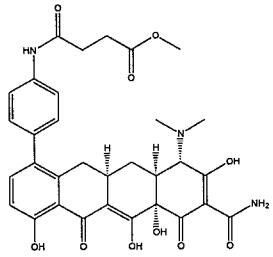
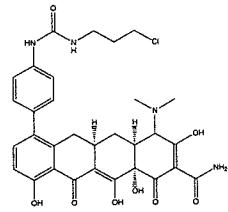
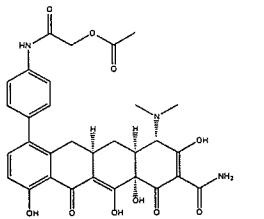


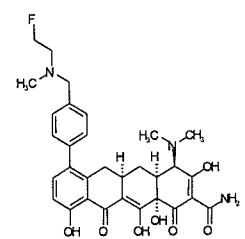
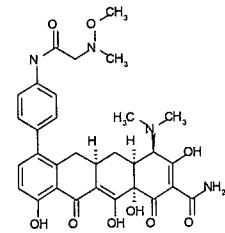
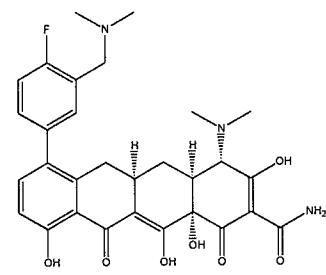
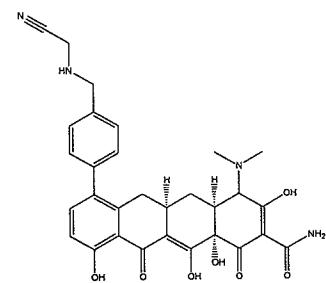
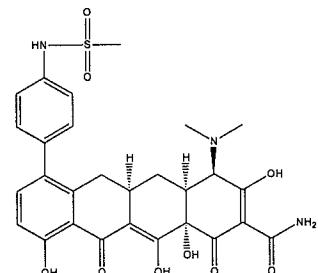
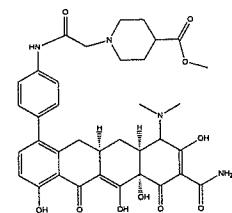
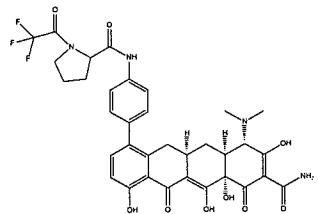
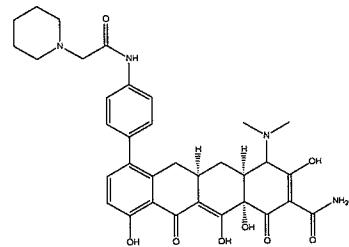
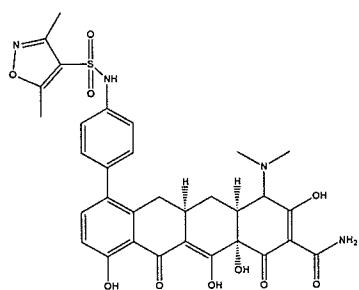


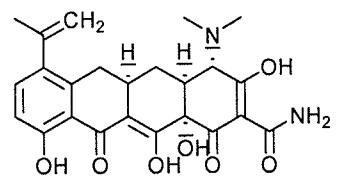
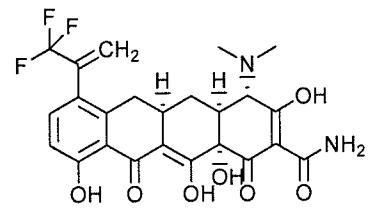
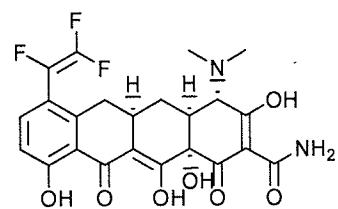
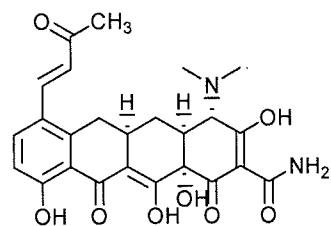
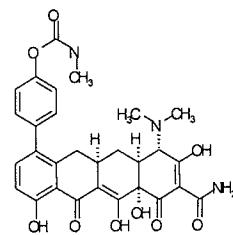
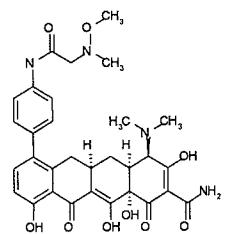
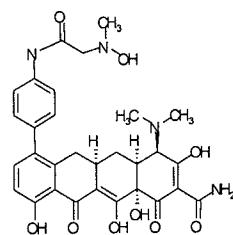
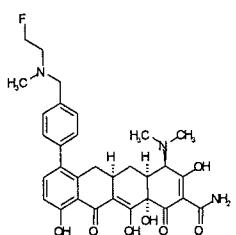
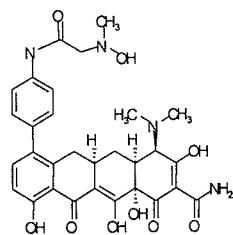
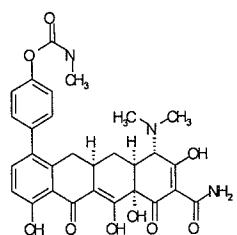


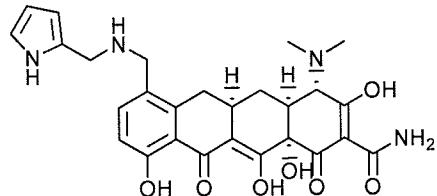
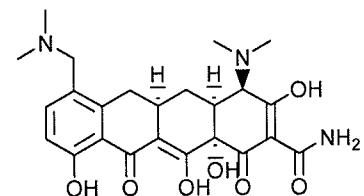
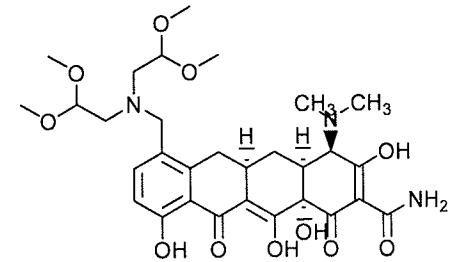
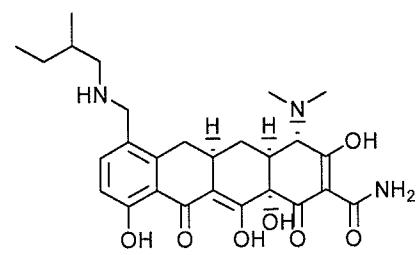
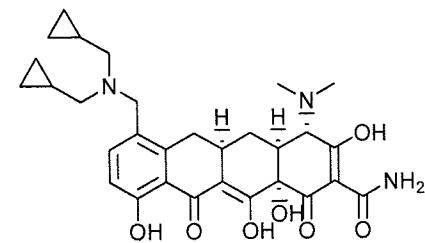
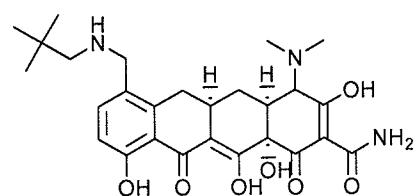
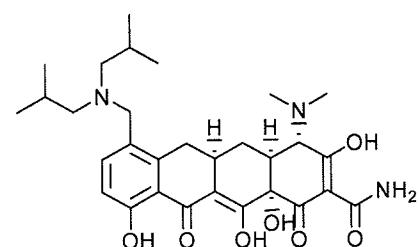
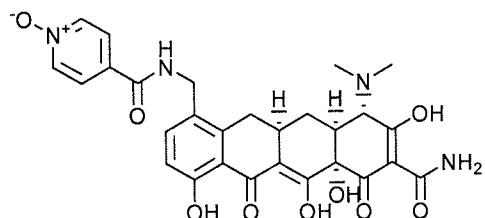
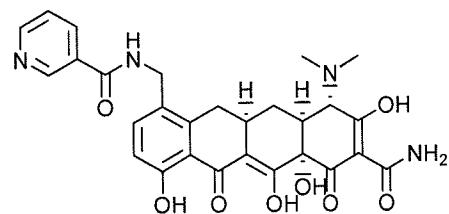
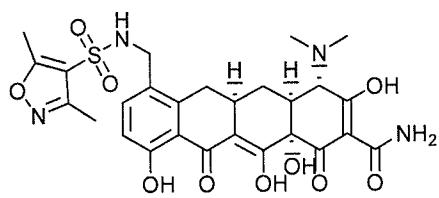


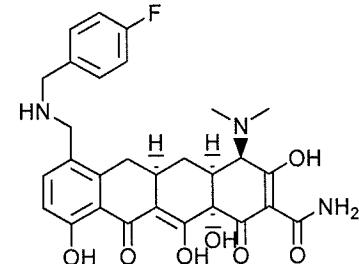
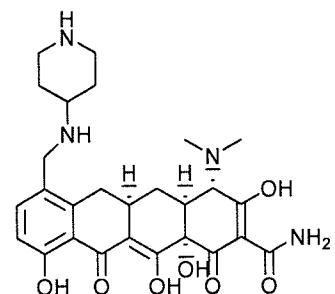
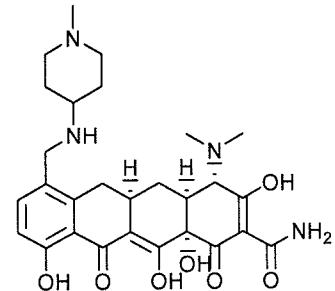
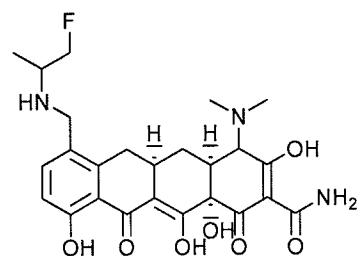
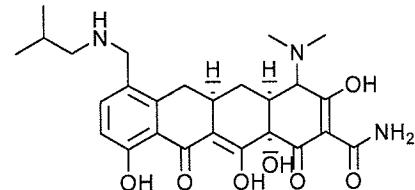
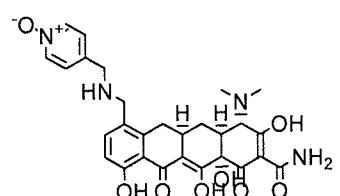
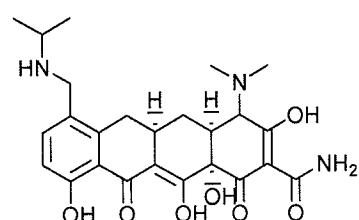
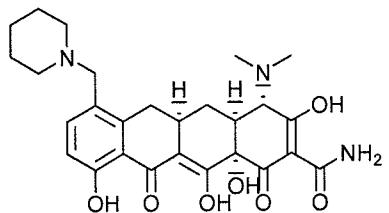
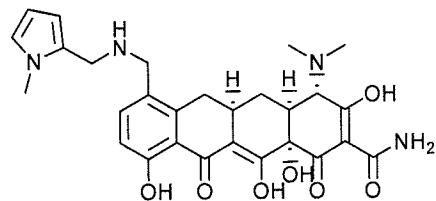
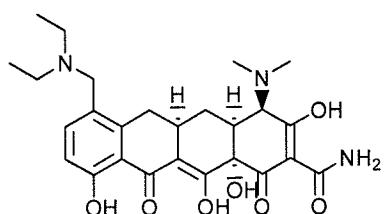


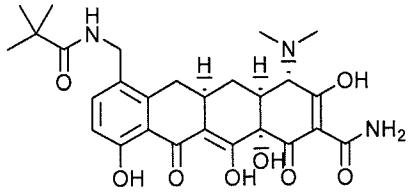
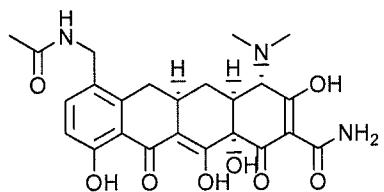
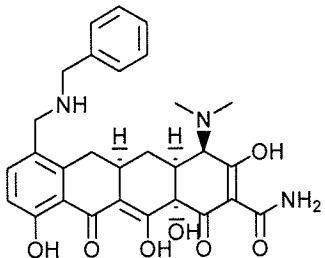
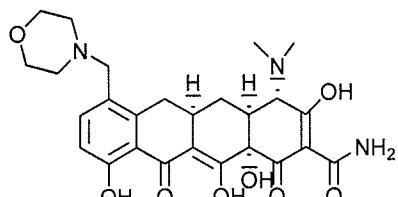












and pharmaceutically acceptable salts, esters and enantiomers thereof; such that said DTMR associated with splicing of nuclear RNA is treated, wherein said DTMR associated with splicing of nuclear RNA is spinal muscular atrophy, and further wherein said effective amount is effective to modulate splicing of said subject's nuclear RNA.